

SAMUEL BERKOWITZ
EDITOR

Cleft Lip and Palate

*Diagnosis
and Management
2nd Edition*



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Editor **Cleft Lip and Palate**
Samuel Berkowitz

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Cleft Lip and Palate

2nd Edition

With 478 Figures, Mostly in Color,
and 46 Tables

 Springer

Editor

SAMUEL BERKOWITZ, DDS, MS, FICD
Diplomate, American Board of Orthodontics
Maxillo-Mandibular Reconstruction
Cranio Facial Orthopedics
The Professional Center – Suite 112
6601 S.W. 80 St.
South Miami, FL 33143, USA

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Dedication

My professional growth has been nurtured by my understanding wife, Lynn, who made it possible for me to spend endless uninterrupted evenings at my desk, while at the same time encouraging me to “stay with it.” Warm hugs to my two daughters, Beth and Debra, Ruben and Edward, and my eight grandchildren for their endless expressions of support and love.

Last, but by no means least, I cannot say enough for the countless children with various palatal and facial clefts whom I have treated over the past four decades and for their understanding parents. This book is ded-

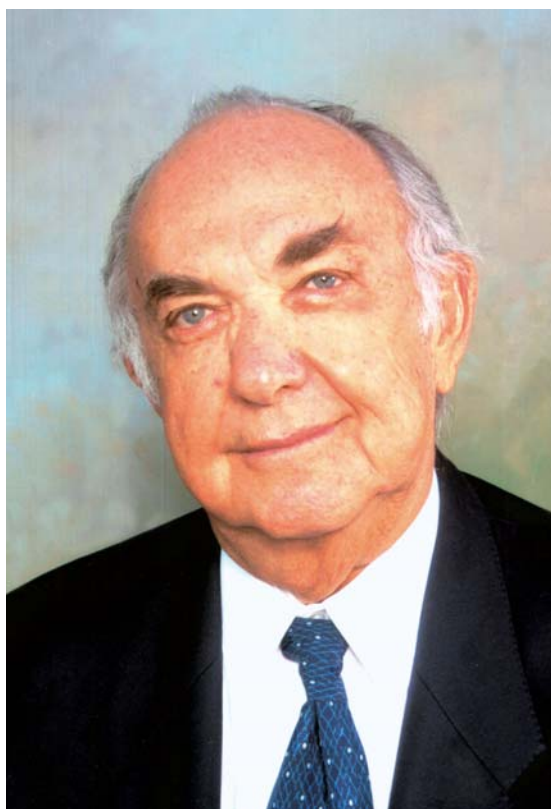
icated to all of them as a token of my appreciation for their enduring perseverance and fortitude. My young patients have taught me much about the human spirit and the joy that can spring from surmounting nature’s adversities.

Finally, my work was made possible by the support of J. Ralph Millard Jr., who appreciated the value of serial records starting at birth. He and I have differed on a few areas of treatment, but we strongly agreed that only through the analyses of objective growth records could progress in treatment be accomplished.

About the Editor

Dr. Berkowitz, an orthodontist, was a Clinical Professor of Pediatrics and Surgery associated with the South Florida Craniofacial Anomalies Program at the University of Miami School of Medicine. Currently he is Adjunct Clinical Professor at Nova Southeastern University College of Dentistry – Orthodontic Department, and Adjunct Clinical Professor of Orthodontics at the University of Illinois College of Dentistry. His main goal is to develop teaching materials in cleft palate for professionals in plastic and oral surgery, orthodontics, and speech language pathology. He is a past President of the American Cleft Palate Association Educational Foundation, and the Florida Cleft Palate Association, and is currently President of the Miami Craniofacial Anomalies Foundation. Dr. Berkowitz was active in the American Association of Orthodontics, Florida Cleft Palate Association, and The Edward Angle Society of Orthodontists. He has published widely in medical and cleft palate journals and is the author of Volume I and the editor of Volume II of *Cleft Lip and Palate Perspectives In Management* – First Edition; he coauthored *Plastic Surgery of the Facial Skeleton* with S.A Wolfe, M.D, and wrote *The Cleft Palate Story* for parents of a child born with a cleft. Dr. Berkowitz is a popular speaker on cleft lip/palate topics and has presented many workshops and seminars in the USA and abroad.

His research interest focuses on improving surgical-orthodontic treatment planning for cleft lip and palate children as well as those with other craniofacial anomalies. Currently, Dr. Berkowitz is project director of a clinical research program that is studying the long-term effects of various surgical treatment procedures on palatal and facial growth and development. He created a quantitative method for determining when to close the palatal cleft space, based on the 10% ratio of the cleft space to the area of the surrounding palatal surface medial to the alveolar ridges. He is creating a Power-Point lecture series for surgeons and



Samuel Berkowitz, DDS, MS, FICD

orthodontists to enable them to better understand and teach others the effects of surgery on the face from birth through adolescence.

Dr. Berkowitz has been awarded the title “Honoree” by the Edward Angle Society of Orthodontists, and “Honoree” by the First World Congress of the

International Lip and Palate Foundation for his many contributions to the field of cleft lip and palate treatment. His extensive serial clinical records of dental casts, lateral cephaloradiographs, facial and intraoral photographs, and panorex are going to the National

Museum of Health and Medicine (associated with Walter Reed Hospital's Institute of Pathology in Washington D.C), where they will be available for continued study.

Foreword

It is most gratifying to be able to write a foreword to this latest and most valuable addition to our compendium of knowledge about cleft lip and palate. The field has been close to my heart for over 50 years, even before I became Director of Research at Northwestern University's Cleft Lip and Palate Institute in 1948. It has been my good fortune to be associated with some of the outstanding pioneers in the Team Effort approach – Herbert Cooper, Wayne Slaughter, Sam Pruzansky, J. Daniel Subtelny, Howard Aduss, Jack Thompson, Alan Brodie, Herbert Koepp-Baker, Harold Westlake, Fred Merrifield, Wilton Marion Krogman, Sam Berkowitz, Robert Ricketts, Margaret Hotz, Rudi Hotz, Arnold Huddart, Sheldon Rosenstein, Bengt Johansson, Hans Friede, Mohammed Mazaheri, Karin Vargervik, Samir Bishara, Donald Warren, Hughlett Morris, Morten Rosen, Charles Kremenak, Bill Olin, Ralph Millard Jr., Ralph Shelton, Ken Salyer, and many others in the U.S. and Europe. These dedicated and knowledgeable leaders in the field built a strong foundation of total service for patients unfortunate enough to develop this congenital defect.

My own research in the growth and developmental aspects and the influence of therapeutic ministrations has been replicated and serves to remind us of the complexities of the biologic continuum and their interrelationships. My maxim always has been, "From the abnormal, we learn much about the normal."

Samuel Berkowitz wrote his master's thesis in cleft palate under the supervision and guidance of Samuel Pruzansky at the Craniofacial Program at the Univer-

sity of Illinois School of Dentistry in 1959. From there he went to the University of Miami School of Medicine to help develop, with Dr. D. Ralph Millard Jr., Chief of Plastic Surgery, a craniofacial anomalies program and clinic (1960–1998). They collaborated in developing an extensive collection of longitudinal records of dental casts cephaloradiographs, panorex, and photographs from birth to adolescence. Dr. Berkowitz's main goal was to create lasting treatment concepts based on a better understanding of the natural history of cleft palate and facial growth and development. This book discusses in detail the resulting treatment concepts, which are supported by in-depth case analyses.

Dr. Berkowitz has drawn on the experience an international array of scholars and practitioners – researchers, surgeons, orthodontists, speech therapists, pediatricians, obstetricians, psychologists, prosthodontists, pediatric dentists, otolaryngologists, audiologists, and others. He has carefully crafted and integrated the important contributions from each field, welding these diverse areas into a multidisciplinary team. These are described in the preface. There is no doubt in my mind that this work will become the standard reference for all who work in the field of craniofacial anomalies, as we move into the twenty-first century.

T. M. Graber, DMD, MSD, PhD, Odont. Dr., DSc
Editor of the International Journal
of Orthodontics and Dentofacial Orthopedics

Preface

In the first page of the first edition of this book, I quoted Samuel Pruzansky [1] who, after participating at an International Symposium on Cleft Lip and Palate held in 1969, and reflecting on what he heard at that meeting, stated, “The same tired questions have been asked as at every similar clinical meeting. And I despair at the general unfamiliarity with the pertinent literature.”

Fortunately, since the 1950s, many clinical investigators in the field of cleft palate have performed excellent clinical studies of the management of cleft lip and palate that have contributed to the intellectual ferment over the last 50 years. To these studies we are indebted, since to know this literature is vital for correct treatment planning.

When selecting significant references for this text, every attempt was made to carry out an exhaustive literature search to include all of the excellent articles on each subject covered. That, however, has been an insurmountable task. To investigators whose research articles were not included, I apologize and I advise readers to conduct their own literature search, which must include papers on the “opposing schools” of thought. There is no doubt in my mind that their final conclusions will be the same as mine when they consider the results of long-term palatal and facial growth studies that involved the analysis of objective records.

To familiarize clinicians with the appropriate literature and its importance to the treatment of cleft lip and cleft palate, the chapters in this book are structured to improve clinicians’ understanding of the natural history of the cleft defect, the face in which it exists, the influence of surgery on palatal growth and development, and equally importantly in developing an appreciation for the heterogeneity that exists even within a single cleft type.

These chapters will show that chronological age is not the parameter that really matters in determining the age at which to close the cleft in the palate. What is

important is morphologic age and physiologic fitness, that is, whether the tissues are adequate in quantity and quality and whether the geometric relationship of cleft parts is favorable or unfavorable for reconstruction. Some questions incident to growth, which date back 25 years, concern the relationship of the malformed palatal segments to the contiguous skeletal anatomy, which, in turn, may be anomalous. These following questions are also addressed: Are the palatal segments static in their deficiency or does the deficiency diminish in time, that is, is “catch-up-growth” a predictable phenomenon? And if so, what surgical procedures (as to age and type) make it possible?

Many of Pruzansky’s thoughts, written so many years ago, still hold true today and are worth repeating. He stated that *whoever sees things from their beginning will have the most advantageous view of them*. To that end, most of the serial cases presented in this volume start soon after birth when plaster casts and photographs of the palatal and facial defect are taken. Serial lateral cephaloradiographs are added as soon as the child is manageable, and again taken periodically through adolescence.

It is hoped that clinicians who are just beginning their involvement in cleft palate will learn the pathology and its natural history of cleft palate from the cases presented in this book and appreciate the need to keep careful records (casts, cephaloradiographs, photographs, and panorex) which are of vital importance to both the processing of knowledge and self-criticism.

One last note of great importance – it is rare that two members of a team, such as I, an orthodontist, and D. Ralph Millard Jr., a plastic surgeon, can successfully work together even when some differences in treatment philosophy exist. We succeeded because we were professionally compatible and because we shared an obsessive need to determine why some procedures are successful and why others fail even when the same treatment procedures were used. Failures, we discov-

ered, occur principally because of misinterpretation of physiological principles and/or a lack of technical proficiency.

Dr. Millard understands the value of serial objective records dating from birth as the essential starting point in determining the long-term utility of any surgical cleft treatment program. Although I was always free to voice a contrary opinion as to what surgery should be performed (and when), our working relationship was based on recognizing the right of the surgeon to reject recommendations and follow his own dictates. And it was my right, as a member of a team involved in growth studies, to document the anatomical changes to the face and palate for future analysis. Respecting our mutual rights and responsibilities was no simple task. Strong emotional and conceptual barriers had to be overcome in the process of communicating with each other.

Our 40-year search for a better understanding of the natural history of cleft lip/palate growth and development and the effects of various surgical-orthodontic treatment procedures ultimately led Dr. Millard to a conservative approach of staged surgical treatment without the intercession of maxillary orthopedics with periosteoplasty, which he tried and found wanting.

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Special appreciation is extended to those who attended my Cleft Palate Seminar and contributed financial support to the Miami Craniofacial Anomalies Foundation.

Immeasurable thanks are likewise due to my many colleagues in the American Cleft Palate-Craniofacial Association and involved in various cleft palate clinics in Europe and Asia for contributing to my understanding of cleft lip and palate management. To them, too many to recognize by name, I shall be forever grateful for their professional knowledge and personal friendship.

Introduction

The general aim of this volume is to present recognized experts from the clinical sciences of dentistry, medicine, speech, audiology, psychology, genetics, ethics, and biology, so that all aspects of the treatment of cleft palate and other craniofacial anomalies can be scrutinized from a particular point of view: long-term clinical experience.

For the sake of brevity, many variations in cleft type and their treatment alluded to in this book were not presented. Because of the multiplicity of variables, no simple description or classification and treatment plan could possibly satisfy everyone concerned with this problem.

Pruzansky [1] was once asked, "When should the orthodontist's, speech pathologist's, or prosthodontist's interest in the cleft palate child begin?" His response: "The answer is quite clear. Everyone who seeks to serve the needs of the child with a cleft should begin at the beginning". An interest in all events affecting these children is essential to the training and educational experience that each member of the team must obtain. Each specialist emerges not only better informed in his own field, but with an increased perspective regarding the means available for providing an integrated program of care for the handicapped child.

The material presented examines the face with a cleft in all aspects as a biologic continuum from birth through postnatal growth and development to maturity at various stages of treatment. In the past several decades, many advances have taken place in cleft habilitation procedures. Unfortunately, many of these changes have not fulfilled all of their stated objectives, and in some instances, these procedures were found to be either injurious or at best unnecessary. These errors will be discussed in detail.

This book also brings together clinicians and biological scientists from the United States, Asia, and

Europe, each of whom in his or her own way has been seeking answers to the multifaceted problem of cleft palate, regarding its embryopathogenesis, craniofacial growth, maxillary orthopedics, surgery, protraction of the maxilla, dental speech prostheses, secondary alveolar bone grafting, speech, hearing, genetics, psychosocial development, and craniofacial surgery.

Each contributor presents pertinent concepts so that a broad perspective of the entire habilitative process can be obtained. The conclusions the reader will reach will be the result of well-documented literature of selected well-controlled clinical research that has withstood the test of review and re-examination.

Because space limitations prevent thorough penetration of all aspects of each subject, a large bibliography is included for additional source material. In no way could these chapters be expected to cover all aspects of this complex subject.

It is my hope that, through a better understanding of the cleft palate defect and face, all clinicians will be better able to evaluate present-day treatment practices and concepts to better plan their own treatment procedures.

We fully acknowledge the important contributions made by the authors and research programs from the institutions which have strongly influenced much of what has been written in these volumes

All lip and palate surgery of my cases were performed by Dr. Ralph Millard, Jr., except where otherwise indicated; S.A. Wolfe performed all skeletal surgery and secondary alveolar bone grafting. They both performed superior-based pharyngeal flaps. No presurgical orthopedics were used unless specifically indicated.

Samuel Berkowitz, MS, DDS, FICD
Editor

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Contents

I	Facial Embryology and Neonatal Palatal Cleft Morphology	
1	Developmental Biology and Morphogenesis of the Face, Lip and Palate	
	<i>Alphonse R. Burdi</i>	
1.1	Summary	9
	References	10
2	Prenatal Diagnosis of Oral Clefts	
	<i>Jorge L. Gomez</i>	
	References	15
3	The Value of Longitudinal Facial and Dental Casts Records in Clinical Research and Treatment Analysis	
	<i>Samuel Berkowitz</i>	
3.1	Serial Cephaloradiographs and Casts of the Maxillary and Mandibular Dentition and Occlusion	17
3.2	The Beginning of Longitudinal Cleft Palate Research Studies	20
3.3	Research Methods	20
3.3.1	Retrospective Studies	20
3.3.2	Prospective Studies	20
3.3.3	Clinical Trials	20
3.3.3.1	Randomization of Surgical Procedures	20
3.3.3.2	The Ethics of Surgical Retrospective Clinical Trials (RCT)	21
3.3.3.3	Informed Consent	21
3.4	The Need for Geometric and Quantitative Analysis of Cleft Palate Casts	21
	References	22
4	Facial and Palatal Growth	
	<i>Samuel Berkowitz</i>	
4.1	Maxillary and Mandibular Growth Concepts	23
4.1.1	Newborn Palate with a Cleft of the Lip or Palate	23
4.1.2	Genetic Control Theory: Craniofacial Growth is Entirely Predetermined	23
4.1.3	Functional Matrix Theory	23
4.1.4	Cartilage-Directed Growth: Nasal Septum Theory	25
4.1.4.1	Stimulation of Bone Growth – Is it Possible?	26
4.1.4.2	The Need to Prevent Collapse	26
4.1.5	Basion Horizontal Concept: The Direction of Facial Growth	28
4.2	Mandibular Development in Cleft Palate	30
4.3	Patterns of Postnatal Growth	31
4.3.1	Bone Remodeling During Growth	32
4.3.2	Maxillary Growth	33
	References	33

5	Alternative Method Used to Correct Distorted Neonatal Cleft Arch Forms	
	<i>Samuel Berkowitz</i>	
5.1	Effects of Reversing the Facial Force Diagram	35
5.2	Variations in the Palate's Arch Form	35
5.3	Reversing Aberrant Cleft Facial Forces in the Neonate	35
5.3.1	Lip Surgery, Elastic Traction, or Presurgical Orthodontic Treatment	35
	References	40

II Types of Clefts

6	The Effect of Clefting of the Lip and Palate on the Palatal Arch Form	
	<i>Samuel Berkowitz</i>	
6.1	Varieties of Cleft Lip and Cleft Palate	43
6.1.1	Action of Intact Facial Muscular Forces on the Maxillary Arch	43
6.2	Aberrant Muscle Forces in Clefts of the Lip and Palate	43
6.3	Categories of Clefts	45
6.3.1	Clefts of the Lip	46
6.3.2	Cleft Lip and Cleft Palate	48
6.3.3	Isolated Cleft Palate	50
6.3.4	Submucous Cleft Palate	52
6.4	Congenital Palatal Insufficiency (CPI)	53
	References	53
6A	Clefts of the Lip and Alveolus and Clefts of the Uvulae and Soft Palate	
	<i>Samuel Berkowitz</i>	
6A.1	Clefts of the Lip and Alveolus	55
6A.2	Clefts of the Uvulae and Soft Palate and Cleft of the Uvulae Alone	60
6B	Complete Unilateral Cleft of the Lip and Palate	
	<i>Samuel Berkowitz</i>	
6B.1	Facial Characteristics	63
6B1.1	The Oslo Study	64
6B1.2	Multicenter CUCLP Cephaloradiographic Study	64
6B1.3	Reflection on Ross' Excellent Multicenter Study	65

6B.2	How the Palate Grows	66
6B.3	Treatment Sequence	66
6B3.1	Usual Treatment Sequence	66
6B.4	Reports	66
	References	98
6C	Complete Bilateral Cleft Lip and Palate	
	<i>Samuel Berkowitz</i>	
6C.1	Premaxillary Protrusion: Real or Apparent. Is the Palate Deficient in Bone?	99
6C.2	The Premaxillary-Vomerine Suture	102
6C.3	Facial Growth Studies Show That Midfacial Retrusion Is Not Predictable	105
6C.4	Long-Term Facial Growth Findings Show Class III Outcomes Are Not Predictable	105
6C.5	The Vomer Flap: Good or Bad? Are all Vomer Flaps the Same?	108
6C.5.1	External Elastics Attached to a Head Bonnet or Elastic Tape Strapped to the Cheeks	108
6C.5.2	Uniting the Lip	109
6C.6	Profile Changes	109
6C.6.1	Why Some Premaxillae Continue to Project Following Lip Repair and Others Do Not	113
6C.6.2	Dental Occlusion	113
6C.6.2.1	After Birth	113
6C.6.2.2	In the Deciduous Dentition (3–6 Years of Age)	118
6C.6.2.3	Mixed Dentition (6–11 Years of Age)	118
6C.6.2.4	At Adolescence	118
6C.6.2.5	Retention	119
6C.7	The Following Case Studies Represent Conservative Surgical and Orthodontic Treatment Sequence	119
	References	190
6D	Isolated Cleft Palate	
	<i>Samuel Berkowitz</i>	
6E	Submucous Cleft Palate	
	<i>Samuel Berkowitz</i>	

7 Lip Pits; Orthodontic Treatment, Dentition and Occlusion; Associated Skeletal Structures

Samuel Berkowitz

7.1	Lip Pits	201
7.1.1	Pits of the Lower Lip in Cleft Lip and/or Palate – Genetic Considerations	201
7.1.2	Frequency	201
7.1.3	Morphology	202
7.1.4	Association with Other Malformations	203
7.1.5	Inheritance	204
7.1.6	Evidence of Heterogeneity	204
7.2	Orthodontic Treatment, Dentition and Occlusion	204
7.2.1	Crossbite Correction	204
7.2.1.1	Unilateral Cleft Lip and Palate	207
7.2.1.2	Bilateral Cleft Lip and Palate	208
7.2.1.3	Use of Orthopedic Forces to Correct Midfacial Recession	208
7.2.2	Supernumerary (Extra) Teeth, Missing Teeth, and Aplasia (Malformed Teeth)	208
7.2.3	Caries	209
7.3	The Relationship Between the Clefting Process and Contiguous Skeletal Structures ...	210
7.3.1	The Position of the Cleft Maxilla Within the Cranium and the Mandible	210
7.3.2	The Cranial Base	210
7.3.3	Relationship of the Nasal Cavity to Arch Form	211
	References	212
8	Pierre Robin Sequence	
	<i>Samuel Pruzansky, Julius B. Richmond</i>	
8.1	Growth of Mandible in Infants with Micrognathia	215
8.1.1	Case 1	216
8.1.2	Case 2	218
8.1.3	Case 3	219
8.2	Comment	221
8.3	Summary and Conclusions	222
	References	222

III Facial Growth in Cleft Palate Children

9 Characteristics of Facial Morphology and Growth in Infants with Clefts

Sven Kreiborg, Nuno V. Hermann, Tron A. Darvann

9.1	Introduction	225
9.2	The Danish Experience	225
9.2.1	Cleft Lip (CL)	227
9.2.2	Cleft Palate (CP)	227
9.2.3	Robin Sequence (RS)	227
9.2.4	Cleft Lip and Palate (CLP)	229
9.3	Discussion and Conclusions	229
9.3.1	Intuitive Visualization of the Location of Growth Differences	230
	References	234

10 Facial Growth and Morphology in the Unoperated Cleft Lip and Palate Subject: The Sri Lanka Study

Michael Mars

10.1	Sri Lankan Cleft Lip and Palate Project .	237
10.1.1.	Controls	238
10.1.2	Records Collected for Study	238
10.1.3	Radiographs	238
10.2	Unoperated Unilateral Cleft Lip and Palate	241
10.2.1	Dental Study Models	241
10.2.2	The GOSLON Yardstick	245
10.2.3	Unilateral Cleft Lip and Palate Study Models Analysis by Reflex Microscope	245
10.2.3.1	Arch Widths	245
10.2.3.2	Tooth Widths	245
10.2.3.3	Chord Lengths	246
10.2.3.4	Crossbites	246
10.2.3.5	Overjet	246
10.2.3.6	Missing Teeth	246
10.2.3.7	Crowding	246
10.2.4	Summary of Reflex Microscope Findings on Study Models: UCLP	247

10.3	Unoperated Bilateral Cleft Lip and Palate	247
10.4	Unoperated Isolated Clefts of the Palate	249
10.4.1	Isolated Cleft Palate Study Models Analysis by Reflex Microscope	249
10.4.1.1	Tooth Sizes	249
10.4.1.2	Chord Lengths and Perpendicular Distances	249
10.4.1.3	Arch Widths	249
10.4.1.4	Overjets	249
10.4.2	Factors Influencing Interpretation of Results from the Sri Lankan Cleft Lip and Palate Project	251
10.4.2.1	Malnutrition and Growth	251
10.4.2.2	Speech Implications	252
10.4.2.3	Racial Variation	252
10.4.2.4	Surgical Implications	254
	References	254
11A	A Brief Overview of Psychological Issues in Cleft Lip and Palate <i>Kathleen A. Kapp-Simon</i>	
11A.1	Social and Emotional Adjustment	257
11A.2	Cognitive Development and School Achievement	258
11A.3	Summary	259
	References	259
11B	Craniofacial Psychology: New Directions <i>Joyce M. Tobiasen</i>	
11B.1	Stigma, Self Concept, and Social Psychological Adjustment .	264
11B.2	Self-Protective Properties of Social Stigma	264
11B.3	Research on Self-Protective Properties of Facial Deformity	265
11B.3.1	Attributing Negative Feedback to Prejudice	265
11B.3.2	Selective Comparisons to Similar Groups	266
11B.3.3	Selectivity of Values	266
11B.4	Surgery and Self-Esteem	267
11B.5	Research and Clinical Implications	268
	References	268

IV Lip and Palate Surgery

12	A Short History of Prepalatal Clefts <i>P. Randall, D. LaRossa</i>	
	References	280
13	Core Curriculum for Cleft Lip/Palate and other Craniofacial Anomalies	
14	Palatal Wound Healing: The Effects of Scarring on Growth <i>Johannes W. Von den Hoff, Jaap C. Maltha, Anne Marie Kuijpers-Jagtman</i>	
14.1	Introduction	301
14.2	Wound Healing	301
14.2.1	Skin and Oral Mucosa	301
14.2.2	Phases in Wound Healing	302
14.2.3	Contraction and Scarring	303
14.3	Effects of Palatal Repair on Growth ...	304
14.4	Experimental Research	305
14.4.1	Effects of Surgery on Growth	305
14.4.2	Modification of Surgical Techniques ..	306
14.4.3	Tissue Engineering	306
14.4.3.1	Biocompatible Membranes	307
14.4.3.2	Epithelial Sheets	307
14.4.3.3	Composite Substitutes	307
14.4.4	Mechanisms of Wound Healing (In Vivo Studies)	308
14.4.5	Mechanisms of Wound Healing (In Vitro Studies)	309
14.5	Application of Experimental Results ..	310
	References	310
15	Lip and Palate Surgery <i>Samuel Berkowitz</i>	
15.1	The Influence of Surgery on Growth ...	315
15.2	Surgical Closure of the Cleft Lip and Palate	316
15.3	Lip Surgery	317
15.3.1	Lip Adhesion	317
15.4	Palatal Cleft Surgery: Type, Timing, and Sequence	319
15.4.1	What to Do and When to Do It: Speech and Palatal Growth Considerations	319

15.4.1.1	False Premise 1/2 Wrong Conclusions 1/2 Therapeutic Folly	319
15.5	The Effect of Surgery on Maxillary Growth	324
15.6	Speech Considerations	326
15.7	Surgical-Orthodontic Procedures and Sequences	327
15.7.1	Palate Cleft Closure Controversies Revisited	328
15.7.2	Scarring Inhibits Palatal Growth	328
15.8	Dental Occlusion Associated with Early Palatoplasty Using a Vomer Flap	341
15.8.1	The Fourth Dimension of Time: Catch-up Growth	342
15.8.2	The Need for Differential Diagnosis . .	342
15.8.3	Timing of Palatal Closure Based on the Ratio of the Palatal Cleft to the Palatal Size	343
15.9	Good Speech Is Dependent on a Palate of Relatively Normal Size and Shape	344
15.10	Facial Changes in Successfully Treated Cases	351
15.10.1	Lateral Cephalometric Results from the Oslo Team	351
	References	351
16	Diseases of the Ear in Children with Cleft Palate and Craniofacial Anomalies <i>Sylvan E. Stool</i>	
16.1	The Auditory System	355
16.1.1	Observations of the Tympanic Membrane	355
16.1.2	Eustachian Tube	357
16.1.3	Eustachian Tube Function Test	359
16.2	The Sequelae of Middle-Ear Effusion in the Cleft Palate Child	361
16.3	Craniofacial Anomalies and Communication	361
	References	362

17	Timing of Cleft Palate Closure Should Be Based on the Ratio of the Area of the Cleft to That of the Palatal Segments and Not on the Age Alone <i>Samuel Berkowitz</i>	
17.1	Method and Material	363
17.1.1	Method Used for Analyses	364
17.2	Treatment Protocols at Each of the Centers	365
17.2.1	Miami Craniofacial Anomalies Foundation, South Florida Cleft Palate Clinic	365
17.3	Results – Comparison of Total Surface Area in Unilateral Cases	367
17.3.1	Growth Velocity in the Unilateral Cases	368
17.3.2	Comparison of Unilateral Posterior Cleft Areas	370
17.3.3	Comparisons of the Ratio of Posterior Cleft Area to Total Surface Area in Unilateral Cases	370
17.3.3.1	Tracking of the Large and Small Segments in Unilateral Cases	370
17.3.4	Comparisons of Surface Area in the Bilateral Cases	372
17.3.4.1	Growth Velocity in the Bilateral Cases	373
17.3.5	Comparison of Bilateral Posterior Cleft Areas	373
17.3.6	Comparisons of the Ratio of Posterior to Total Surface Area in Bilateral Cases	373
17.3.7	Conclusions for the Bilateral Series . .	374
17.3.8	Clinical Significance of the Results . .	374
17.3.9	Discussion	375
17.4	Good Speech Is Dependent on a Normal Palate	376
17.5	Conclusions	377
	References	377

V Presurgical Orthopedics

18 Neonatal Maxillary Orthopedics

Samuel Berkowitz

18.1	The Beginning	381
18.1.1	Stated Benefits of Presurgical Palatal Manipulation ...	381
18.2	Closing the Alveolar Cleft Space: Primary Bone Grafting	382
18.2.1	The Kernahan-Rosenstein Procedure .	383
18.2.2	Critical Review	383
18.2.3	Long-term Results	383
18.2.4	A Critique of Primary Bone Grafts ...	384
18.3	The Zurich Concept	385
18.4	The Netherlands Approach	385
18.4.1	Unilateral Cleft Lip and Palate (UCLP)	385
18.4.2	Bilateral Cleft Lip and Palate (BCLP) ..	387
18.4.2.1	The Spread of PSOT Clinics	390
18.4.2.2	Long-term Utility of PSOT	390
18.4.3	The Long-term Effect of Primary Bone Grafting	391
	References	392
19	History of Neonatal Maxillary Orthopedics: Past to Present	
	<i>Anne Marie Kuijpers-Jagtman, Birte Prah-Andersen</i>	
19.1	Introduction	395
19.2	Early History of Neonatal Maxillary Orthopedics	396
19.3	Claimed Benefits of Neonatal Maxillary Orthopedics	396
19.4	Specific Types of Infant Orthopedics	397
19.4.1	Kernahan Rosenstein Procedure	397
19.4.2	Latham-Millard Pinned Appliance	398
19.4.3	The Zurich Approach	398
19.4.4	Nasoalveolar Molding Grayson	399
19.5	The DUTHCLEFT Study	400
19.5.1	Background	400
19.5.2	Experimental Design	400
19.5.3	General Effects	401
19.5.4	Orthodontic Effects	401
19.5.5	Effect on Speech	402
19.5.6	Cost-Effectiveness	403
19.6	Conclusions	404
	References	404

20

A Comparison of the Effects of the Latham-Millard POPLA Procedure with a Conservative Treatment Approach on Dental Occlusion and Facial Aesthetics in CUCLP and CBCLP

Samuel Berkowitz, Martha Mejia

20.1	Dental Occlusion	409
20.1.1	Method and Materials	409
20.1.1.1	POPLA – Presurgical Orthopedics with Lip Adhesion	410
20.1.1.2	Non-POPLA Conservative Treatment of CBCLP and CUCLP	414
20.2	Discussion	414
20.3	POPLA CUCLP	418
20.4	Conservative Non-POPLA CUCLP and CBCLP Cases	444
20.5	Variations in Palatal Osteogenic Deficiency and Its Influences on Surgical Treatment	448
20.6	Correction of Midfacial Deficiencies in Conservatively Treated Non-POPLA Cases	448
20.7	Similar Presurgical Orthopedics As It Was Utilized in the Past – It Failed Then As It Does Now	448
	References	449

21 Nasoalveolar Molding for Infants Born with Clefts of the Lip, Alveolus and Palate

Barry H. Grayson, Deirdre Maull

21.1	Introduction	451
21.2	History	451
21.2.1	Objectives	452
21.3	Procedure	453
21.4	Complications	456
21.5	Benefits	457
	References	458

22 Surgical Treatment of Clefts of the Lip and Palate from Birth to Age Ten

*S. Anthony Wolfe, Rami Ghurani,
Marta Mejia*

22.1	Timing of Treatment	459
22.2	Presurgical Treatment	460
22.3	Palatal Closure	461

22.4	Secondary Repair of Palatal Defects	462
22.5	Alveolar Bone Grafting	462
22.6	Secondary Operations on the Lip and Nose	463
22.7	Case Reports	463
	References	476

VI Midfacial Changes

23A	Protraction Facial Mask <i>Samuel Berkowitz</i>	
23A.1	Protraction of the Maxilla Using Orthopedics	479
	References	485
23B	Protraction Facial Mask for the Correction of Midfacial Retrusion: The Bergen Rationale <i>Rolf S. Tindlund</i>	
23B.1	Early Rehabilitation	487
23B.2	Midfacial Retrusion in CLP Patients	487
23B.2.1	Anterior Crossbite	488
23B.2.2	Orofacial Function	488
23B.3	Principles of Orthopedic/Orthodontic Treatment in CLP Patients	488
23B.3.1	Checklist for CLP Orthopedic/Orthodontic Treatment Objectives ..	489
23B.3.1.1	Presurgical Orthopedics	489
23B.3.1.2	Interceptive Orthopedics	489
23B.3.1.3	Alignment of Maxillary Incisors	489
23B.3.1.4	Secondary Alveolar Bone Grafting ...	489
23B.3.1.5	Conventional Orthodontics in the Permanent Dentition	489
23B.3.1.6	Dental Adjustments at Age 16–17 for Girls, 18–19 years for Boys	495
23B.4	Outline of CLP Treatment Procedures in Bergen	495
23B.4.1	Plastic Surgery	495
23B.4.2	Interceptive Orthopedics	495
23B.4.2.1	Protraction Facial Mask	495
23B.4.2.2	Quad-helix Spring (with Four Bands and Hooks)	495
23B.4.2.3	Transverse Expansion	497
23B.4.2.4	Protraction	498
23B.4.2.5	Fixed Retention	498

23B.4.3	Treatment Results of Using a Protraction Facial Mask ..	498
23B.4.3.1	Clinical Results	498
23B.4.3.2	Limitations	499
23B.4.3.3	Stability/Relapse	499
23B.4.3.4	Soft-Tissue Profile	499
23B.4.4	Long-Term Prognosis After Interceptive Orthopedics	500
23B.5	Conclusions	500
	References	501

23C LeFort I Osteotomy

S.A Wolfe, Samuel Berkowitz

23C.1	Surgical Maxillary Advancement LeFort I Osteotomy	503
23C.2	Stability of Maxillary Advancement	505
23C.3	Total Maxillary Advancement and Its Possible Effect on Speech	510
23C.4	Technique	510
23C.5	Multiple Maxillary Osteotomies	515
	References	516

23D Rigid External Distraction: Its Application in Cleft Maxillary Deformities

John W. Polley, Alvaro A. Figueroa

23D.1	Materials and Methods	519
23D.2	Cephalometric Evaluation	521
23D.3	Results	522
23D.3.1	Angular Changes	522
23D.3.2	Linear Changes	523
23D.3.3	Dental Changes	523
23D.4	Discussion	524
	References	531

VII Orthognathic Surgery

24 Management of Maxillary Deformities in Growing Cleft Patients

Eric J.W. Liou, Philip K.T. Chen

24.1	Introduction	535
24.2	Orthopedic Management of Hypoplastic Maxilla in Growing Unilateral or Bilateral Cleft Patients ..	535
24.2.1	Effective Maxillary Orthopedic Protraction	535

24.2.1.1	Double-Hinged Rapid Maxillary Expander	536	24.5.3.5	Postdistraction Maintenance and Orthodontic Tooth Movement Through the Regenerate	551
24.2.1.2	Alternate Rapid Maxillary Expansions and Constrictions (Alt-RAMEC) of Maxilla	537	24.5.3.6	Postdistraction Alveolar Bone Grafting or Gingivoperiosteoplasty ...	551
24.2.1.3	Maxillary Protraction Springs for Effective Maxillary Orthopedic Protraction	538	24.5.3.7	Treatment Results	551
24.2.1.4	Treatment Protocol for Effective Maxillary Orthopedic Protraction	538	24.6	Summary	553
24.2.1.5	Treatment Results and Effects of Effective Maxillary Orthopedic Protraction	538		References	553
24.3	Orthopedic Management of a Downward Displaced Premaxilla in Bilateral Cleft Patients	540	25	Remodeling the Craniofacial Skeleton by Distraction Osteogenesis – The Madible	
24.3.1	Premaxillary Orthopedic Intrusion ...	541		<i>Fernando Molina</i>	
24.3.1.1	Orthodontic Preparation	541	25.1	Clinical Applications of Distraction Osteogenesis	555
24.3.1.2	Device for Premaxillary Orthopedic Intrusion	541	25.1.1	Hemifacial Microsomia	555
24.3.1.3	Treatment Results of Premaxillary Orthopedic Intrusion	541	25.1.2	Micrognathias	561
24.3.1.4	Mechanisms of Premaxillary Orthopedic Intrusion	543	25.1.3	The Temporomandibular Joint Ankylosis	563
24.4	Orthopedic Management of a Laterally Displaced Premaxilla in Bilateral Cleft Patients	544	25.1.4	Cleft Lip and Palate Patients	563
24.4.1	Premaxillary Orthopedic Medial Repositioning	544	25.1.5	Craniosynostosis	565
24.4.1.1	Orthodontic Preparation	545	25.2	Discussion	569
24.4.1.2	Device for Premaxillary Orthopedic Medial Repositioning	545		References	571
24.4.1.3	Treatment Results of Premaxillary Orthopedic Medial Repositioning	545	26	Cleft-Orthognathic Surgery	
24.4.1.4	Mechanisms of Premaxillary Orthopedic Medial Repositioning	545		<i>Jeffrey C. Posnick, Paul S. Tiwana</i>	
24.5	Managements of a Wide Alveolar Cleft and Fistula	547	26.1	Introduction	573
24.5.1	Protocol for Approximating a Wide Alveolar Cleft	547	26.2	Integrated Team Approach	573
24.5.2	Minimize Alveolar Cleft by Effective Maxillary Orthopedic Protraction	547	26.3	Timing	573
24.5.2.1	Clinical Procedures for Effective Maxillary Orthopedic Protraction of Lateral Segments of Maxilla	547	26.4	Residual Deformities	575
24.5.2.2	Treatment Results	547	26.5	Historical Perspective and Results	575
24.5.3	Interdental Distraction Osteogenesis for Approximating Alveolar Cleft Wider than a Tooth Width	548	26.6	Controversies	580
24.5.3.1	Presurgical Orthodontic Preparations	548	26.7	Conclusions	583
24.5.3.2	Interdental Distraction Site	549		References	583
24.5.3.3	Surgical Procedures	549	27	Prevention of Relapse Following Cleftal Bone Grafting and the Future Use of BMP Cytokines to Regenerate Osseous Clefts Without Grafting	
24.5.3.4	Distraction Protocol	550		<i>Philip J. Boyne, Alan S. Herford, Dale E. Stringer</i>	
			27.1	Iliac Crest Bone Grafting	588
			27.2	Appropriate Sequencing of Orthodontic and Surgical Treatment	589
			27.3	Description of the Surgical Procedure for Bone Grafting	592

27.4	Relapse in Bilateral Cases Following Bone Grafting and Possible Causes of Relapse	595
27.5	The "Displaced" Premaxilla	595
27.6	The Use of Different Types of Bone Graft Materials	596
27.7	Surgical Use of BMP Cytokines in Producing Appropriate Reconstruction of Cleftal Bone Defects	596
27.8	Review of Studies of Cytokines in Bone Inductive Reconstructive Surgery (BMPS)	598
27.9	Application of BMP-2 to Maxillary Cleft Defects	598
27.10	Conclusions	599
27.11	Summary	600
	References	600
28	Secondary Bone Grafting of Alveolar Clefts <i>Frank E. Abyholm</i>	
28.1	Surgical Technique	601
28.2	Orthodontic Management	602
28.2.1	Preparation for Bone Grafting	602
28.2.2	Permanent Dentition Management ...	602
28.3	Optimal Age for Secondary Bone Grafting	603
28.3.1	Secondary Bone Grafting	604
28.4	The Grafting Tissue of Choice	604
28.4.1	Donor Sites	604
28.4.2	Complications	604
28.5	Important Surgical Details	605
28.5.1	Flap Design	605
28.5.2	Good Exposure	605
28.5.3	Cancellous Bone Only	605
28.6	Conclusion	605
	References	606
29	Speech Implication of Orthognathic Intervention <i>Donna Russell Fox</i>	
29.1	Role of Speech Language Pathologist	607
29.2	Effects of Altering the Vocal Tract on Speech	607

29.3	Preoperative Considerations for Orthognathic Surgeons	610
	References	610

VIII The Nasopharyngeal Area

30	Diagnostic Procedures and Instruments Used in the Assessment and Treatment of Speech <i>Samuel Berkowitz</i>	
30.1	Articulation Tests	615
30.2	Rating Scales of Speech Intelligibility and Acceptability	615
30.3	Cephalometrics	615
30.4	Cine- and Videofluoroscopy	616
30.5	Multiview Videofluoroscopy	616
30.5.1	Technique	617
30.6	Ultrasound	617
30.7	Video Nasopharyngoscopy	617
30.7.1	Technique	618
30.8	The Nasometer	618
30.8.1	Technique	619
30.9	Aeromechanical Measurement	619
30.9.1	Warren and Dubois Technique	619
30.9.1.1	PERCI	619
30.9.1.2	TONAR	619
30.10	Summary	619
	References	619
31	Variations in Nasopharyngeal Skeletal Architecture <i>Samuel Berkowitz</i>	
31.1	Muscles	621
31.1.1	Pharynx and Velum	621
31.2	Nasopharyngeal Growth	622
31.3	Functions	623
31.3.1	Swallowing	623
31.3.2	Speech	623
31.4	The Role of the Nasal Cavity	625
31.5	The Use of Lateral Roentgen-cephalometrics in Evaluating Skeletal Pharyngeal Architecture and Velar Elevation	627
31.6	Cervical Spine Anomalies	628
31.7	Velar Closure	632

31.8	Improving Velopharyngeal Closure ...	635	33.5	Effects of Maxillary Expansion on the Nasal Airway	662
31.8.1	Pharyngeal Flaps	635	33.6	Effects of the Nasal Airway on Speech	663
31.8.2	Speech Aid Appliances	637		References	664
31.8.3	Push-Back Procedures (Velar Lengthening)	640	34	Surgical Management of Velopharyngeal Dysfunction	
31.8.4	Sphincteric Pharyngoplasty (SP) of Orticochea	640		<i>Richard E. Kirschner, Rachel A. Ruotolo</i>	
31.8.5	Posterior Pharyngeal Wall Augmentation	641	34.1	Patient Evaluation	667
	References	641	34.2	Posterior Pharyngeal Flap	669
32	The Velopharyngeal Mechanism		34.2.1	Surgical Technique	670
	<i>Robert J. Shprintzen</i>		34.2.2	Outcome	671
32.1	Normal Velopharyngeal Closure	643	34.3	Sphincter Pharyngoplasty	671
32.2	Failures of Velopharyngeal Valving ...	646	34.3.1	Surgical Technique	672
32.2.1	Disorders of Structure	646	34.3.2	Outcome	672
32.2.1.1	Overt Clefts	646	34.3.3	Comparison of Posterior Pharyngeal Flap and Sphincter Pharyngoplasty ...	673
32.2.1.2	Submucous Cleft Palate	648	34.4	Furlow Double Opposing Z-Palatoplasty	673
32.2.1.3	Occult Submucous Cleft Palate	648	34.4.1	Surgical Technique	674
32.2.1.4	Other Structural Disorders	649	34.4.2	Outcome	675
32.3	Neurogenic and Myopathic Disorders of Velopharyngeal Function	650	34.5	Posterior Pharyngeal Wall Augmentation	675
32.4	Evaluation of Velopharyngeal Insufficiency	650	34.5.1	Surgical Technique	676
32.5	Indirect Assessments of VPI	650	34.5.2	Outcome	676
32.6	Direct Assessments	651	34.6	Management of Persistent VPD	677
32.6.1	Multiview Videofluoroscopy	651	34.6	Summary	678
32.6.2	Nasopharyngoscopy	653		References	678
32.7	Which Direct Visualization Procedure to Use?	654	35	Velopharyngeal Dysfunction Management Algorithms	
32.7.1	Multiview Videofluoroscopy Advantages	654		<i>Jeffrey L. Marsh</i>	
32.7.2	Multiview Videofluoroscopy Disadvantages	654	35.1	Evaluation of Velopharyngeal Function	682
32.7.3	Nasopharyngoscopy Advantages	654	35.2	Differential Diagnosis of Velopharyngeal Dysfunction	683
32.7.4	Nasopharyngoscopy Disadvantages ...	654	35.3	Differential Management of Velopharyngeal Dysfunction	684
32.8	Reporting of the Results	654	35.3.1	Small Midline Gap	685
32.9	The Importance of It All	654	35.3.2	Sagittal Gap with Active to Moderately Active Lateral Pharyngeal Wall Motion	685
	References	655	35.3.3	Hypodynamic or Adynamic Velopharyngeal Sphincter	685
33	The Nasal Airway in Breathing and Speech		35.4	Conclusion	686
	<i>Donald W. Warren, Amelia F. Drake</i>			References	686
33.1	Children	658			
33.2	Adults	660			
33.3	Effects of Secondary Procedures on the Nasal Airway	661			
33.4	Speech Aid Prostheses and the Nasal Airway	661			

IX Speech

36	Optimal Age for Palatoplasty to Facilitate Normal Speech Development: What is the Evidence?	
	<i>Sally Peterson-Falzone</i>	
36.1	Why Is It So Difficult to Answer this Question?	691
36.2	Why Do Speech-language Pathologists Worry So Much About Early Development?	692
36.3	What Constitutes Scientific Evidence in Clinical Research?	692
36.4	A General Warning: "A" Does Not Cause "B"	695
36.5	A Review of the Literature up to 2000	695
36.6	The Ongoing Dilemma of "Primary Veloplasty"	697
36.7	A Summary of What We Have Learned Since 2000	700
36.8	Conclusion	700
	References	700
37	Speech, Language, and Velopharyngeal Dysfunction: Management Throughout the Life of an Individual with Cleft Palate	
	<i>John E. Riski</i>	
37.1	Introduction	705
37.2	Challenges	705
37.3	Predicting the Need for Secondary Palate for Management	706
37.4	Predicting Future Speech Proficiency	707
37.5	Articulation Development and the Age at Palatoplasty	708
37.6	The Stability of Velopharyngeal Functioning of Time	708
37.7	Age at Pharyngoplasty	709
37.8	The Adult Patient	710
37.9	Primary Versus Secondary Pharyngoplasty	710
37.10	Speech Therapy	711
37.11	Role of Parents in Speech Therapy	712
37.12	Language Development and Learning	712
37.13	Preschool and School-Aged Investigations	714
37.14	Velo-Cardio-Facial Syndrome	715
37.15	Summary	716
	References	717
38	Prosthetic Speech Appliances for Patients with Cleft Palate	
	<i>Mohammed Mazaheri</i>	
38.1	Diagnosis and Treatment Planning	721
38.2	Treatment Planning	721
38.3	Requirements of a Speech Appliance	721
38.4	Indications for Prostheses in Unoperated Palates	722
38.4.1	A Wide Cleft with a Deficient Soft Palate	723
38.4.2	A Wide Cleft of the Hard Palate	723
38.4.3	Neuromuscular Deficiency of the Soft Palate and Pharynx	723
38.4.4	Delayed Surgery	724
38.4.5	Expansion Prosthesis to Improve Spatial Relations	724
38.4.6	Combined Prosthesis and Orthodontic Appliance	724
38.5	Indications for a Prosthesis in Operated Palates	726
38.5.1	Incompetent Palato-pharyngeal Mechanisms	726
38.5.2	Surgical Failures	726
38.6	Contraindications for a Prosthesis	726
38.7	Constructing Prosthetic Speech Appliances	726
38.7.1	Preliminary Impression	727
38.7.2	Preparation of the Deciduous Teeth for Retention	727
38.7.3	Final Impression	728
38.7.4	Jaw Relation Records	728
38.8	Design and Construction of the Prosthesis	729
38.8.1	Construction of Velar Section	729
38.8.2	Construction of Pharyngeal Section or Speech Bulb	729
38.8.3	Insertion of the Appliance	732
38.8.4	Position of Speech Bulb	732
38.9	Summary	732
	References	733

39	Palatal Lift Prosthesis for the Treatment of Velopharyngeal Incompetency and Insufficiency <i>Mohammed Mazaheri</i>		
39.1	Treatment, Methodology, and Results in Patients with Velopharyngeal Inadequacy	735	
39.1.1	The Referral	735	
39.1.2	Results of Treatment	735	
39.1.3	Summary	739	
39.1.4	Conclusion	739	
39.2	Palatal Lift Prostheses for the Treatment of Patients Requiring Velar Elevation, Velopharyngeal Stimulation, and Velopharyngeal Obturation	739	
39.2.1	Symptoms	739	
39.2.2	Etiology	739	
39.2.3	Speech Characteristics	740	
39.2.4	Methods of Treatment	740	
39.2.5	Prerequisites of Lift and Combination Prostheses	740	
39.2.6	Objectives in Making Prosthetic Lift and Combination Services	741	
39.2.6.1	Results of Using Lift and Combination Prostheses	741	
39.3	Summary	745	
	References	746	
X The Future			
40	Summary of Treatment Concepts and a New Direction for Future Palatal Growth Studies <i>Samuel Berkowitz</i>		
40.1	A New Direction for Cleft Research	751	
40.2	Clinical Research	751	
40.2.1	Initial State	751	
40.2.2	Maneuver: Presurgical Orthopedics and Surgical Procedures Used to Close the Palatal Cleft	752	
40.3	Palatal Embryopathology	752	
40.4	The Neonatal Palatal Form in Complete Clefts of the Lip and Palate	753	
40.4.1.	The Effect of Muscle Forces	753	
40.4.2	The Influence of Cleft Surgery on Palatal Form and Growth	753	
40.5	The Need for Three-Dimensional Measuring Techniques	754	
40.6	Studies Using Three-Dimensional Techniques	755	
40.6.1	Study 1: Analysis of Longitudinal Growth of CUCLP and CBCLP	755	
40.6.1.1	Statistical Methods	755	
40.6.1.2	Results	761	
40.6.1.3	Discussion	762	
40.6.2	Study 2: Quantitative Study of a Patient with CBCLP Treated with Presurgical Orthopedics	762	
40.7	A New Instrument for 3D Facial and Palatal Cast Study	762	
	References	762	
41	Eurocleft – An Experiment in Intercenter Collaboration <i>W.C. Shaw, G. Semb</i>		
41.1	The Eurocleft Cohort Study	765	
41.1.1	Outcomes at Age 9	766	
41.1.2	Follow-up	766	
41.1.3	Survey of Treatment Experience	766	
41.1.4	Consistency of Outcomes Over Time	766	
41.1.5	Lack of Association Between Outcome and the Amount of Treatment	767	
41.1.6	Lack of Association Between Outcome and Satisfaction	768	
41.1.7	The Benefits of Intercenter Comparison	768	
41.1.8	Audit vs. Research	769	
41.1.9	Alternatives to Intercenter Comparisons for Routine Clinical Audit	769	
41.1.9.1	Good Practice Archive	769	
41.1.9.2	Registries	770	
41.1.9.3	Benchmarks	770	
41.1.10	International Agreement on Record Collection and Comparison Methodology	770	
41.2	Eurocleft Clinical Network	771	
41.2.1	Creation of the Network	771	
41.2.2	Development of Consensus Recommendations	771	
41.2.3	Survey of European Services	771	
41.2.3.1	Models of Care and National Policies	771	
41.2.3.2	Clinical Practices	772	
41.2.3.3	Consumer Associations in Europe	773	
41.2.4	The Challenge of Improving Quality of Services	773	
41.2.5	Subsequent Progress	773	

41.3	EUROCRAN	773	42	Social, Ethical, and Health Policy Issues in the Care of Children with Major Craniofacial Conditions	
41.3.1.1	A Group of Randomized Control Trials of Primary Cleft Surgery	774		<i>Ronald P. Strauss</i>	
41.3.1.2	A Prospective Registry of Patients Undergoing Distraction Osteogenesis	774	42.1	Introduction	777
41.3.1.3	Two Cohort Studies of Surgical Outcome in UCLP	774	42.2	Craniofacial Treatment Decision-Making	777
41.3.1.4	Development of a Good Practice Archive	774	42.2.1	The History of Care for Children with Major Craniofacial Conditions ..	778
41.3.1.5	A Gene-Environment Interaction Study	774	42.2.2	The Gatekeeper Role for Physicians ...	778
41.3.1.6	A Chromosomal Approach to Identifying Orofacial Clefting Genes	774	42.2.3	The Impact of Prenatal Diagnosis	780
41.3.1.7	Molecular Diagnosis of Monogenic Craniofacial Anomalies	775	42.2.4	Social Justice and Resource Allocation	780
41.3.2	The Global Context	775	42.3	Justice and the Distribution of Resources	782
	References	775		References	782
				Subject Index	785

Contributors

FRANK E. ABYHOLM, MD, DDS
Dept. of Plastic Surgery
Rikshospitalet National Hospital
Sognsvannsvn 20, Oslo, 0027, Norway

SAMUEL BERKOWITZ, DDS, MS, FICD
Diplomate, American Board of Orthodontics
Maxillo-Mandibular Reconstruction
Cranio Facial Orthopedics
The Professional Center – Suite 112
6601 S.W. 80 St.
South Miami, FL 33143, USA

PHILIP J. BOYNE, DMD, MS, DSc
Department of OMFS
Loma Linda University School
of Dentistry, Loma Linda, CA 92350, USA

ALPHONSE R. BURDI, PhD, SciD
University of Michigan
Dept. of Cell & Developmental Biology
5734 Medical Science Bldg. 2
Ann Arbor, MI 48109-0616, USA

PHILIP K.T. CHEN, MD
Dept. of Orthodontics
and Craniofacial Dentistry
Chang Gung Memorial Hospital
199 Tung-Hwa North Road, Taipei, 105, Taiwan

TRON A. DARVANN, MSc, PhD
The 3D-Laboratory for Image Processing
and Visualization, School of Dentistry
Faculty of Health Sciences
University of Copenhagen
Copenhagen University Hospital (Rigshospitalet)
and the Technical University of Denmark
Copenhagen, Denmark

AMELIA F. DRAKE, MD
Craniofacial Outcomes Registry
University of North Carolina at Chapel Hill
1700 Airport Road, CB# 5136
Chapel Hill, NC 27599-5136, USA

ALVARO A. FIGUEROA, DDS, MS
Rush Craniofacial Center
Rush University Medical Center
Professional Bldg. I, Suite 425
1725 West Harrison Street, Chicago, IL 60612, USA

RAMI GHURANI, MD, DDS
Craniofacial Fellow
Miami Children's Hospital
W-3100 SW 62nd Ave., Miami, 33155, USA

JORGE L. GOMEZ, MD
7300 SW 62nd Place, Suite 201, Miami, FL 33143, USA

BARRY H. GRAYSON, DDS
New York University Medical Center
550 First Avenue, New York, NY 10016, USA

ALAN S. HERFORD, DDS, MD
Chairman, Division of Oral & Maxillofacial Surgery
Dept. of Surgery
Loma Linda University Medical Center
Loma Linda, CA 92350, USA

NUNO V. HERMANN, DDS, PhD
Assistant Professor
Dept. of Pediatric Dentistry and Clinical Genetics
School of Dentistry, Faculty of Health Sciences
University of Copenhagen, Nørre Allé 20
2200 Copenhagen, Denmark

KATHLEEN A. KAPP-SIMON, PhD
Cleft Lip & Palate Institute
2225 Enterprise Dr., Se. 2506A
Westchester, IL 60154-5805, USA

RICHARD E. KIRSCHNER, MD, FACS, FAAP
Children's Hospital of Philadelphia
34th & Civic Center Blvd., Wood Bldg. 1st Floor
Philadelphia, PA 19104, USA

SVEN KREIBORG, DDS, PhD
School of Dentistry, Faculty of Health Sciences
University of Copenhagen, Nørre Allé 20
2200 Copenhagen, Denmark

ANNE-MARIE KUIJPERS-JAGTMAN
University of Nijmegen
Dept. of Orthodontics and Oral Biology
6500 HB Nijmegen, The Netherlands

D. LAROSSA
3400 Spruce Street, Philadelphia, PA 19104, USA

ERIC JEIN-WEIN LIOU, DDS, MS
Dept. of Orthodontics and Craniofacial Dentistry
Chang Gung Memorial Hospital
199 Tung-Hwa North Road, Taipei, 105, Taiwan

DEIRDRE MAULL, DMD
Inova Fairfax Hospital for Children
Craniofacial Program, Annandale, VA 22003, USA

JAAP C. MALTHA
University of Nijmegen
Dept. of Orthodontics
P.O. Box 9101, 6500 HB Nijmegen, The Netherlands

MICHAEL MARS, BDS, PhD, DSc
Hospital For Sick Children
Maxillofacial & Dental Dept.
Great Ormond Street, London, WC1N 3JH, UK

JEFFREY L. MARSH, MD
Kids Plastic Surgery
777 S. New Ballas Rd., Ste. 300E
St. Louis, MO 63141, USA

MOHAMMAD MAZAHERI, MDD, DDS, MSc
223 North Lime St., Lancaster, PA 17602, USA

MARTA MEJIA, DDS
Research Associate, Division of Plastic Surgery
Miami Children's Hospital
W-3100 SW 62nd Ave, Miami, FL 33155, USA

FERNANDO MOLINA, MD
Hospital Angeles del Pedregal
Camino a Santa Teresa # 1055
PB 06, Colonia Heroes de Padierna
Mexico City, 10700, Mexico

SALLY J. PETERSON-FALZONE, PhD
5460 West Sharpshooter Court
Tucson, AZ 85743, USA

JOHN W. POLLEY, MD
John W. Curtin Chair
Dept. of Plastic & Reconstructive Surgery
Co-Director, Rush Craniofacial Center
Rush University Medical Center, Professional Bldg. I
Ste. 425, 1725 West Harrison Street
Chicago, IL 60612, USA

JEFFREY C. POSNICK, DMD, MD
5530 Wisconsin Ave., Ste. 1250
Chevy Chase, MD 20815, USA

BIRTE PRAHL ANDERSEN
Academic Centre for Dentistry Amsterdam
Louwesweg 1, 1066 EA Amsterdam, Netherlands

SAMUEL PRUZANSKY, †

PETER RANDALL, MD
609 Foulkeways, Gwynedd, PA 19436, USA

JULIUS B. RICHMOND, MD
(address is unknown)

JOHN E. RISKI, MS, PhD
Children's Healthcare of Atlanta
Ste. 420, 5455 Meridian Mark Rd., NE
Atlanta, GA 30342, USA

RACHEL A. RUOTOLO, MD
Children's Hospital of Philadelphia
34th & Civic Center Blvd., Wood Bldg., 1st Floor
Philadelphia, PA 19104, USA

DONNA RUSSELL FOX, MA, PhD
Speech Pathologist
Cronin and Brauer Facial Reconstruction Ins.
Professor Emeritus, University of Houston
8966 Chatsworth, Houston, TX 77024, USA

GUNVOR SEMB
The University of Manchester
School of Dentistry
Higher Cambridge Street, Manchester, M15 6FH, UK

WILLIAM C. SHAW
The University of Manchester
School of Dentistry
Higher Cambridge Street, M15 6FH, Manchester, UK

ROBERT J. SHPRINTZEN, MS, PhD
Center for the Diagnosis, Treatment
& Study of Velo-Cardio-Facial Syndrome
Center for Genetic Communicative Disorders
Communication Disorder Unit
Professor of Otolaryngology
Professor of Pediatrics
Upstate Medical University
750 East Adams Street, Jacobsen Hall 714
Syracuse, NY 13210, USA

SYLVAN E. STOOL, MD
1056 East 19th Ave., B-455
Denver, CO 80218, USA

RONALD P. STRAUSS, DMD, PhD
The University of North Carolina at Chapel Hill
UNC School of Dentistry, CB # 7450
378 Dental Office Building
Chapel Hill, NC 27599-7450, USA

DALE E. STRINGER, DDS
Associate Professor, Dept. of Surgery
Division of Oral & Maxillofacial Surgery
Loma Linda University Medical Center
Loma Linda, CA 92350, USA

ROLF S. TINDLUND, DDS, PhD
Dental School, University of Bergen
Dept. of Orthodontics and Facial Orthopedics
Arstadveien 17, Bergen, 5009, Norway

PAUL S. TIWANA DDS, MD, MS
Dept. of Oral and Maxillofacial Surgery
The University of North Carolina at Chapel Hill
Chapel Hill, NC 27599, USA

JOYCE M. TOBIASEN, PhD
One Ward Pkwy., Ste. 107
Kansas City, MO 64112-2117, USA

JOHANNES W. VON DEN HOFF
University of Nijmegen
Dept. of Orthodontics
P.O. Box 9101, 6500 HB Nijmegen, The Netherlands

DONALD W. WARREN, DDS, MS, PhD
UNC Craniofacial Center
University of North Carolina at Chapel Hill
PO Box 1356, Southern Pines, NC 28388, USA

S. ANTHONY WOLFE, MD
1444 NW 14th St., Miami, FL 33125, USA

Developmental Biology and Morphogenesis of the Face, Lip and Palate

ALPHONSE R. BURDI

Prologue. *The “history of man for the nine months preceding his birth would, probably, be far more interesting and contain events of greater moment than all the three score and ten years that follow it.”*

Samuel Taylor Coleridge, *Miscellanies, Aesthetic and Literary*. Circa 1800.

The developmental biology of the face, lip, and palate is best understood against a backdrop of biological paradigms and information drawn from the multidisciplinary worlds of classical embryology, developmental biology, and, today, from the exciting world of molecular biology. The advent of many new and exciting clinical interventional strategies for the treatment of birth defects now allows the clinician to treat the most delicate of craniofacial abnormalities which were beyond the realm of treatment even for the skillful clinician due to lack of appropriate technologies. Even the details of craniofacial morphogenesis, however interesting to the clinician, were also seen as being beyond the everyday clinical realm, or esoteric, prior to the advent of the new wave clinical interventional techniques, such as high resolution imaging and information technologies, in the fields of craniofacial and maxillofacial surgery. Today, there is a rekindled need to understand more of the details of craniofacial morphogenesis, especially as that understanding increases our awareness of the etiology, pathogenesis, and clinical features of a variety of craniofacial defects. The goal of this chapter is to provide a highlighted update or working understanding of the “developmental blueprint” followed in human craniofacial morphogenesis, with a special focus on defects of the face, lip, palate, and associated structures.

While recent advances in developmental and molecular craniofacial biology contribute heavily to the picture of face and palate morphogenesis, there has been an explosion in our fundamental understanding of the very beginnings of these body regions [43]. Such new information clearly centers on the genesis, behavior, and developmental outcomes of many craniofacial “building block” cell types throughout their life span. These fundamental phenomena include patterns of early DNA signaling, gene and biochemical organizers, nuclear and cellular differentiation, proliferation, migration, and, importantly, patterns of inter-

active behaviors at intracellular, cell surface, and extracellular matrix levels. Complete or partial interruptions of any one or combination of these phenomena have been implicated in the identification of etiologic and pathogenic causes of mammalian birth defects, including those of the human craniofacial regions.

Normal and abnormal morphogenesis of the craniofacial regions, and even that of rest of the body, is dependent upon a myriad of cell types and tissues. One of the most important cell types in understanding normal and abnormal craniofacial morphogenesis is the neural crest cell [2, 36]. While the importance of crest cells has been hypothesized for a century or more, not until the advent of neural crest biological markers – first with isotopic labels and later with specific markers as monoclonal antibodies, intracellular dyes, and protein assays – did the neural crest become so widely appreciated in a variety of studies of vertebrate embryogenesis in general, and human craniofacial morphogenesis, in specific. While the majority of recent neural crest studies of have necessarily dealt with chick and mouse embryos, there is ample evidence to show that basic information and associated technologies gained from vertebrate embryos can be directly applied to neural crest cells in mammalian and human embryos.

With this in mind, prime consideration should be given to neural crest cells because they contribute heavily to craniofacial morphogenesis. These important “building-block” cells arise from the final stages in formation of the embryonic neural tube. Neural crest cell specificity is the result of an inductive action by nonneural ectoderm adjacent to the developing neural tube (mediated by bone morphogenetic proteins BMP-4 and BMP-7) on the lateral cells of the neural plate as the plate transforms from a plate of ectoderm into the definitive neural tube. The induced neural crest cells express the transcription factor *slug* which characterizes cells which separate from an em-

bryonic epithelial layer and subsequently migrate as mesenchymal cells away from the parent site [33].

The identification of the exact molecular mechanisms and cellular events linked to the differentiation, proliferation and, and especially, of the migration of crest cells into the facial and pharyngeal regions is not yet fully known. What is known, however, is that, with the variant *OTX2* transcription factor, specific patterns of neural crest proliferation and migration into the pharyngeal arches are controlled by four members of homeodomain proteins called *HOX* genes (A-D). Another factor thought to be critical in the migration of crest cells is a loss of cell-to-cell adhesiveness which is associated with the loss of cell adhesion molecules (CAMs) characteristic of the neural tube and the migrating neural crest cells [91]. Following the completion of craniofacial crest cell migrations and differentiation into specific structures (such as bones of the facial skeleton) CAMs are re-expressed. Migrating craniofacial crest cells are thought to travel through cell-free intercellular spaces and pathways that have high levels of extracellular matrix molecules [3, 9, 10]. These migrations are determined by factors intrinsic to the crest cells themselves and features of the external environment through which the crest cells migrate. While most available information on neural crest migratory pathways comes from chick studies, data suggest that such information is applicable to mammalian and humans as well. Migrations are facilitated by the presence of such molecular substrates as fibronectin, laminin, and type IV collagen. Attachment to and migration of crest cells is mediated by a family of attachment proteins called integrins. It is important to note that other extracellular matrix molecules in the pathway (e.g., chondroitin sulfate-rich proteoglycans) can impede or block the normal migration of neural crest cells, which may lead to a number of craniofacial malformations.

As will be expanded upon later in this chapter, neural crest cells have the remarkable capacity to differentiate into a wide variety of anatomical structures throughout the body. Exactly what controls crest differentiation is still an open question. One hypothesis is that all neural crest cells are equal in their developmental potential, and their ultimate differentiation is entirely predetermined by the environment through which the crest cells migrate and finally reside, i.e., "extrinsic determinants." A second hypothesis favors "intrinsic determinants" and suggests that premigratory crest cells are intrinsically programmed for different developmental fates. Recent studies indicate that both hypotheses may be operative [1]. While neural crest cells have a common site of origin during neural tube formation, not all neural crest cells behave alike. There are two families of crest cells, i.e., cranial and trunk neural crest. Trunk neural crest cells extend

from lower cervical to the most caudal embryonic somites in humans. Trunk neural crest cells appear to have lessened migratory pathways and have fewer developmental outcomes than cranial neural crest cells, including formation of spinal ganglia, sympathetic ganglia, and adrenal medulla chromaffin cells. Interestingly, unlike cranial crest cells, trunk neural crest cells do not have the capacity to differentiate into skeletal tissues.

The life history of cranial neural crest cells, while not of any more importance than trunk neural crest cells, appears to be more complex. Cranial neural crest cells are a major component of the embryo's cephalic end and differentiate into a wide variety of cell and tissue types, including connective, skeletal, dentin, and muscle tissues of the face [64]. Unlike trunk neural crest cells, which show diffuse migratory pathways, cranial neural crest cells follow specific migratory pathways into specific regions of the embryonic head. They arise from the more cephalic neural tube regions and migrate ventrally into the pharyngeal arches adjacent to the upper regions of the embryonic gut tube. Such migrations are extensive and follow very definite migratory paths away from the neural tube and into the facial and pharyngeal regions. In hindbrain regions, neural crest cells arise from eight segmented regions on either side of the hindbrain (rhombencephalon) called rhombomeres (numbered R1-R8) and subsequently migrate into specific pharyngeal arches [12, 14].

Crest cells from R1 and R2 centers migrate into the first pharyngeal arch and play important roles in the formation of Meckel's cartilage and the malleus and incus ear ossicles developing from it. Crest cells from R4 migrate into the second arch and contribute to the formation of the stapes, styloid process, and lesser horn of the hyoid bone. Crest cells from R6 and R7 migrate into the third arch, and those from R8 migrate into the fourth and sixth pharyngeal arches. The one variant noted above is that the first pharyngeal arch also includes crest cells from midbrain levels which express *OTX2* transcription factors. Little evidence exists to show that crest cells from rhombomeric centers R3 and R5 play any significant role in human craniofacial morphogenesis. Crest cells initially express the *HOX* genes from their originating rhombomeric center, but maintenance of a specific expression is dependent upon interaction of the crest cells with the arch-specific mesoderm in the pharyngeal arches. While there is a specific linkage between given *HOX* genes and pharyngeal arches, morphologic derivatives arising from these linkages are also dependent upon epithelial-mesenchymal interactions and includes molecular signaling from surface ectoderm covering the arches, specifically, fibroblast growth factors (FGFs) which interact with underlying mes-

enchymal cells. Crest cells alone do not establish or maintain a specific pattern of morphologic expression. Several regulatory factors have been identified. *Sonic hedgehog* (*Shh*) genes and some retinoids have been shown to regulate normal *HOX* gene expression within the pharyngeal arches associated with a variety of developmental events, including neural plate development and craniocaudal body pattern formation.

Defective differentiation, proliferation, and migration of cranial neural crest have been linked with a variety of developmental defects, i.e., the so-called neurocristopathies [24, 28, 29, 34]. Deficiencies and excesses of retinoids, for example, can disrupt proliferation and migration on specific crest cells, resulting in craniofacial defects, e.g., clefts of the lip and palate [54, 62]. With reference to the structural abnormalities in the chromosome 22-deletion DiGeorge syndrome, the fundamental pathogenesis for this clinical syndrome has been linked with defects in cranial neural crest cells of the third and fourth pharyngeal arches and cardiac outflow tract. Other cranial neurocristopathies include the wide range of craniofacial abnormalities in the frontonasal dysplasia family, Treacher Collins syndrome (mandibulofacial dysostosis), the Robin sequence, Waardenburg syndrome (types I and III), and neurofibromatosis (von Recklinghausen's disease).

The “building block” cells for the head and face are identifiable both premorphologically and morphologically as early as the second intrauterine week. Once mapped out these cells continue with their peak period of cell differentiation, proliferation, and migration through the second intrauterine month. While the classical picture of craniofacial morphogenesis can be framed upon the morphogenesis of primary germ layer cells (i.e., ectoderm, mesoderm, endoderm), there is little doubt whatsoever that the current understandings and excitement about mammalian, including human craniofacial, morphogenesis have been significantly advanced by a plethora of studies of the origins and behavior of embryonic neural crest cells. Morphogenesis of the facial regions depends heavily on the timely differentiation, directed migration, selective proliferation of these crest cells which arise as a product of neural tube formation as the neural tube progressively pinches off from the overlying skin along the body's dorsal axis. As will be discussed later, cells and tissues within each of the embryonic facial primordia arise from neural crest cells begin their migration (at about 21 postconception days) into the facial regions, as cell clusters called rhombomeres, from their sites of origin along the portions of the neural tube which form the brain. The determinants of crest cell migrations have been variously hypothesized as including intrinsic cell “targeting” factors and

chemical signaling from cells lining the extracellular cleavage planes through which the crest cells migrate [37, 42]. Crest cells from the developing midbrain regions migrate into upper facial regions, whereas crest cells from hindbrain migrate selectively into the lower facial regions [45–47]. Importantly, once the crest cells migrate into specific facial regions, they differentiate into mesenchymal cells that subsequently give rise to connective tissue and muscle cells of those specific facial regions [41]. While the predominant neural crest-derived mesenchymal cells in the facial regions do co-mingle with mesodermally derived mesenchymal cells, the interactive nature of their co-mingling, or lack thereof, remains uncertain. Consistent with the tenets of the “developmental field concept” [48] in human morphogenesis, both human and experimental studies generally have hypothesized that significant and early interference with normal differentiation, proliferation, and migration embryonic cells, including especially the craniofacial neural crest cells, can lead to isolated and syndromic craniofacial defects, called neurocristopathies, whose occurrence and severity depend on a combination of environmental and genotypic factors specific to a given dysmorphic or phenotypic trait [46, 47, 52].

Having addressed the general developmental features of the craniofacial “building block” neural crest cells, let's turn our specific attention now to the key events in the shaping of the human face, lip, and palate. When the embryo's cephalo-caudal axis is established at about 14 postconception days, the facial developmental field is one of the first of the head regions to appear [48]. Centrally-located in this region is a discrete bilaminar tissue plate, called the oropharyngeal membrane, whose structure and location marks the junction between the oral ectoderm and the endodermal digestive tube. This membrane progressively degenerates through the normal process of apoptosis or “programmed cell death” which involves increased phagocytic or lysosomal activity along the inner and outer surfaces of the membrane. Once the apoptosis of the oropharyngeal membrane is completed at 4 weeks, there is direct continuity between the spaces of the early oral cavity and the pharyngeal regions of the digestive tube. Only rarely does the oropharyngeal membrane fail to degenerate. Interestingly, a similar ecto-endodermal membrane lies at the depth of a groove which separates the first pharyngeal from the second pharyngeal arch. As will be discussed later, this membrane will have a very different and important developmental fate (i.e., does not undergo apoptosis) than that of the oropharyngeal membrane related to the fact that it has, unlike the oropharyngeal membrane, a layer of mesenchyme interposed between its ecto- and endodermal layers.

Clearly as much developmental “shaping” occurs on the lateral aspects of the young embryo’s head as in its frontal regions. At four weeks, a series of lateral surface elevations, called pharyngeal arches, becomes quite prominent on the lateral side of the head. In fact, the appearance of the embryo’s head region at this time closely resembles the gill slit anatomy seen in a comparably-staged fish embryo; however, unlike in the fishes, the surface gill appearance in human embryos is short-lived, except as noted above, in the case of the development of the ear drum (or tympanum). The pharyngeal arches contribute significantly to the formation of the face, palate, and associated structures. Most congenital malformations of the head and neck have their beginnings during the cellular transformation of the pharyngeal arches into their adult derivatives. For example, branchial cysts and fistulae can occur in those rare instances in which human pharyngeal (or gill) clefts fail to smooth over on the lateral side of the neck. As mentioned earlier, cell masses which contribute to the bulging prominence of the arches are the neural crest cells that have migrated into the pharyngeal arches from specific brain regions, and which eventually differentiate into mesenchymal cells and give rise skeletal and muscular structures specific to a given pharyngeal arch.

The first pair of pharyngeal arches are most important in shaping the human face and associated structures and will receive most attention in this chapter. The first pharyngeal arch, often called the mandibular arch, develops as two elevations around the oral opening which was filled in earlier by the oropharyngeal membrane. The larger and lower regions of this arch form much of the mandibular anatomy and the malleus and incus middle ear ossicles, whereas the smaller and upper regions of the first arch on either side of the oral opening give rise to the anatomy of upper lip, teeth, maxilla, zygomatic bone, and squamous portions of the temporal bone. The second pharyngeal arch is located beneath the first arch and is often called the hyoid arch in that it contributes significantly to the formation of the hyoid bone and one of the three middle ear ossicles, called the stapes. These two pharyngeal arches, like each of the other four pharyngeal arches, are separated from each other by a surface pharyngeal groove which grows inwardly to meet an endodermal-lined outpocketing from the developing pharyngeal region, i.e., the first pharyngeal pouch. As is the case with most pharyngeal grooves and pharyngeal pouches, the contact zone between a pharyngeal groove and a pharyngeal pouch is a bilaminar plate of ectoderm and endoderm which eventually degenerates, again through the process of “programmed cell death” and increased phagocytic activity. In the case of the first arch, however, this bilaminar plate is separated by invading crest-derived mesenchymal cells

which have been linked with the failure of that specific plate to degenerate and persist normally throughout life as the adult eardrum, or tympanum. The elevated margins around the first pharyngeal groove develop through the selective proliferation of mesenchymal cells beneath the skin into six separate mesenchymal swellings, called auricular hillocks. These auricular hillocks progressively (from both the first and second pharyngeal arches) enlarge, migrate, and consolidate through programmed cell activity and eventually give rise to the external ear, or auricle. Failure of the auricular hillocks to develop normally can result in auricles of abnormal size, shape, and position as seen in a variety of isolated and syndromic craniofacial birth defects, e.g., first and second branchial arch syndrome, hemifacial microsomia, and microtia. The complete absence of the auricle (anotia) is a rare event.

To complete this picture of the pharyngeal arches, it is important to note that cells within the arches are supplied by pairs of blood vessels, called aortic arches, that distribute blood from the embryonic heart upward through the tissue of each arch toward the brain and then down to the body [49]. As with the pharyngeal arches themselves, not all of the aortic arches persist in humans. The aortic arches of the third, fourth, and sixth do persist and become greatly modified throughout the embryonic period as they are reconstituted as the common carotid arteries which supply the neck, face, and brain. Especially important in this dynamic development of the craniofacial vasculature is the shifting of the primary arterial supply to the embryonic face prior to, during, and following the formation of the secondary palate. Unlike in the adult, prior to the seventh week, the primary source of blood to both the superficial and deep head tissues is the internal carotid artery and its branches. At about 7–8 weeks when the embryonic palatal shelves are experiencing their most critical stages of elevation and closure, an important shift occurs in the primary blood supply to the face and palatal tissues from the internal carotid to the external carotid arterial system. This transition involves a temporary vascular shunt between internal and external carotid systems provided by the stapedial artery. Failure of either the stapedial artery to form or failure of a complete and timely transition to occur has been hypothesized in identifying the pathogenesis of such conditions as palatal clefting and mandibulofacial dysostosis [51].

Considerably dependent upon the timely set morphogenic events that occur from the time of implantation through the fourth week, the embryonic face continues through its “developmental critical period,” which spans the fifth through seventh intrauterine weeks. It is that time period during which in human craniofacial morphogenesis generally is most suscep-

tible to either known or suspected birth defect-producing agents, or teratogens [66]. Arising from the first pharyngeal arch are four primordial or “building block” tissue masses that surround the large central depression of the primitive oral cavity. Continued morphogenesis of the facial prominences depends heavily upon the continuing migration, proliferation, and differentiation of the neural crest cells, under the direction of developmental morphogens, to a point in time when the facial prominences, or primordia, are clearly identifiable as the single median frontonasal prominence, paired maxillary prominences on either side of the frontonasal process, and two mandibular prominences beneath the oral opening. The shape and size of these prominences as well as development of the specific skeletal and muscular structures of each pharyngeal arch are critically dependent upon the continued viability and differentiation of the neural crest cells which are especially sensitive to teratogens, e.g., cortisone and retinoic acid [50, 61].

Continuing further with our focus on the developmental “blueprint” for the face, specifically the lip, it is important to note that the outcomes of several distinct brain-skin interactions in placode formation are also essential in early facial morphogenesis. By the beginning of the fifth week, oval patches of skin ectoderm lateral to the median frontonasal prominence interact with brain tissue to set off an ecto-ectodermal interaction resulting in the development of the two thickened nasal placodes located at the ventrolateral regions of the frontonasal prominence. Neural crest-derived mesenchymal cells along the margins of the nasal placodes proliferate rapidly to produce horseshoe shaped elevations around the placode, called the medial and lateral nasal prominences, whose continued rapid growth gradually forms the nasal pits, or early nostrils. The forward growth of each lateral nasal process forms the ala of the nose, whereas the medial nasal process contributes to the formation of the nose tip, columella, the philtrum, tuberculum, and frenulum of the upper lip, and the entire primary palate. Through the process of relative growth in this area, the nasal placodes gradually “sink” to the depth of each nasal pit. Failure of the nose to develop completely is associated with failure of both nasal placodes to develop. A second important skin-brain interaction gives rise to localized thickenings of surface ectoderm on each side of the embryo’s head which will form the optic lens, retina, and nerve. Importantly, and as will be discussed later, these eye fields are first located on the lateral aspects of the embryo’s head and progressively migrate to the frontal midline at about the time the facial prominences are consolidating into the complete face [8].

Selective differentiation and proliferation of mesenchymal cells cause the maxillary prominences to

enlarge and migrate medially toward each other and the lateral and medial nasal prominences [12]. This migration is associated not only with patterns of cellular growth within the maxillary prominences, but also with timely migration of the eye fields from the lateral to the frontal regions of the embryo’s face during the fifth through eighth weeks [8]. Disturbances in normal eye field migration have been suggested as one possible cause of median facial clefting and the conditions of hypo- and hypertelorisms. Continued medial migration of the maxillary prominences on both sides also moves the medial nasal prominences toward the midline and each other. By the end of the sixth week, each maxillary prominence blends, or merges, with the lateral nasal prominence along a line which demarcates the future nasolacrimal groove and duct. This event then establishes the continuity between the side of the nose, or alar region, formed by the lateral nasal prominence with the cheek region formed from the maxillary prominence. A combination of reduced cell numbers and abnormal migration of mesenchymal cells can lead to the abnormal merging or consolidation of the maxillary and lateral nasal prominences. Although seen infrequently, this can lead to facial defects involving oblique facial clefts, persistent nasolacrimal grooves, and failure of the nasolacrimal duct to develop.

Between the fourth and eight weeks, the medial nasal prominences merge with each other, small lower portions of the lateral nasal prominences, and with cells in the larger maxillary prominences. This subsurface merging of cells, especially between the medial nasal and maxillary prominences, results in the continuity of upper jaw and lip. As part of this consolidation of the medial nasal and maxillary prominences in upper lip formation, two important morphologic events need to occur. First, there is a deepening and downward growth of the nasal pit toward the oronasal cavity as a blind-ending sac whose floor eventually degenerates through programmed-cell death resulting in the formation of the primitive choanae, which allows a continuity between the spaces of the primitive nasal cavity and the common oronasal cavity. An event occurring concomitantly with nasal pit morphogenesis is the formation of the seam between the intermaxillary segment and the maxillary prominence. As these two segments come together in the sixth week, the developmental surface seam of cells between them also elongates as the nasal pit elongates, deepens, and moves downward. This developmental seam, called the nasal fin, essentially forms the floor of the nasal pit and progressively degenerates by increased activity among phagocytic cells on either side of the seam [67]. Once “programmed cell” death of the nasal fin is essentially completed at about the seventh week, mesenchymal

cells from both the intermaxillary and maxillary prominences intermix, leading to fusion of the upper lip segments into the upper lip and its cupid's bow. The completion of the embryonic lips generally occurs about 1 week earlier than the formation of the palate. Thus, the lips and palate have different "developmental critical periods" and, as such teratogens might affect either the lips or palate separately, or in combination. The intermixture of mesenchymal cells within the consolidated lip segments give rise to connective tissue components and muscle fibers within the orbicularis oris ring of the upper lip. Complete or incomplete failure of the nasal fin to degenerate have been associated with unilateral and bilateral clefts of the upper lip which variously involve abnormalities of the orbicularis oris muscle in terms of the numbers and distribution of its muscle fibers as part of the orbicularis ring.

The incidence of orofacial clefts varies in accordance with the variances reported for differing population groups [22]. Examples of such population-specific differences, called polymorphisms, include cleft lip with or without cleft palate, which is one of the most common craniofacial birth defects in human with a reported incidence as high as 1:1000 in whites and higher in Asian and lower in black populations [13, 38, 56]. Another example of population polymorphisms shows that isolated cleft palate occurs more often in females (67%) than in age-matched males. This gender difference has been associated with a longer period of time for palatal closure in females which essentially increases the time during which female embryos might be affected by palatogenic teratogens [6]. Lateral clefts of the lip may or may not be associated with clefts of the palate. Mesenchymal cell deficiency that results in partial or complete failure of the two medial nasal prominences to consolidate into a philtrum can contribute to the formation of such defects as a bifid nose, or the rare median cleft ("hare lip") of the upper lip, as characteristically seen in the autosomally recessive Mohr syndrome [23, 25, 30].

As the consolidation of facial processes progresses through the embryonic period, crest-derived mesenchymal cells within the maxillary prominences rapidly proliferate and differentiate into tissues which form mesenchymal cell fields from which the muscles of facial expression develop, and whose myofibers are innervated by the cranial nerve to the second arch, i.e., the facial nerve. Similarly, crest-derived mesenchyme in the maxillary and mandibular portions of the first pharyngeal arch differentiate predominately into the muscles of mastication, which are innervated by the trigeminal nerve of the first pharyngeal arch. Cells within the mandibular prominence give rise to muscle and connective tissue structures of the lower lip, chin, and lower cheek regions. With the reshaping

and consolidation of the five major facial prominences, a recognizable human face is evident by the end of the eighth prenatal week [11, 40, 55, 58, 59].

Morphogenesis of the mammalian palate is an even more complex process which depends heavily upon the balance of genetic, hormonal, and various growth factors. As the face nears the completion of its "developmental critical period," lateral palatine processes which form the secondary palate grow out from the walls of the still common oronasal cavity. The "developmental critical period" for the palate is from the end of the sixth week through the eighth intrauterine week, or 1 week longer in duration than that of the lip. These palatine shelves first grow medially, then become oriented inferolaterally to lie on either side of the tongue, which is quite precocious in its own development as a muscle-filled epithelial sac that fills much of the oronasal cavity. Nearing 8 weeks, the vertically oriented palatine shelves are progressively repositioned above the tongue mass. This repositioning of the shelves is thought to involve a combination of concurrent events, including a downward contraction of the tongue, an ameboid-like reshaping of the shelves which gradually places them over the tongue surface, an increase in extracellular shelf "forces" (or shelf fluid turgor) which reposition the shelves in a horizontal position, and a downward repositioning of the lower jaw [7, 18, 19]. In reality, normal or abnormal horizontalization of the palatine shelves is related to a combination of these three events. Palatal shelf elevation begins in the posterior regions of the shelves and depresses the tongue downward and forward and this allows the more anterior regions of the shelves to first contact one another near the posterior edge of the primary palate, or in the region of the future incisive canal [4]. Once the shelves are in a horizontal position, the shelves contact each other, and essentially stick together by a combination of interlocking shelf surface microvilli and a proteoglycans surface coating along the medial epithelial edge (MEE) of each shelf [39]. The elimination of the MEE is crucial for normal morphogenesis of the anterior regions of the secondary palate [15, 20, 26, 32]. Once the shelves make contact, there is a degeneration (i.e., apoptosis or programmed cell death) of epithelial cells along the abutting shelf linings, and a directed movement of crest-derived mesenchymal cells from one shelf to the other. This process of epithelial degeneration along with intershelf bridging of mesenchymal cells is called fusion.

As is the case for craniofacial morphogenesis in general, several categories of factors (e.g., chromosomes, genes, signaling proteins, transcription factors, specific proteins) have either been identified or hypothesized as important in normal and abnormal palatogenesis [57, 64]. Fusion of the palatal shelves

has been linked to a variety of growth factors like those in the TGF- β 3 growth factor family [32, 53, 63] and protein activities [16, 31]. Complete or regional failures in programmed cell death (MEE) along the lengths of the palatal shelves can lead to various forms of palatal clefts. While great strides have been made in identifying genetic, cellular, and molecular controls of normal palatal development (mostly in mice and chick embryos), the identification of the exact balance between intrinsic and environmental controls of palatal morphogenesis remains elusive [44, 60]. The embryonic palatine raphe, or future midpalatine suture, marks the line of fusion between the palatine shelves. From the site of first shelf contact and fusion near the future incisive foramen, fusion of the more posterior regions of the shelves takes place over the next two weeks. Fusion also occurs between the shelves and the inferior edge of the nasal septum, except in the more posterior regions where the soft palate and uvula remain free. Once fusion of the shelves of the secondary palate is complete, their mesenchymal cells differentiate into osteogenic cells which form the skeletal elements of the premaxillary, maxillary, and palatine portions of the palate.

Formation of the soft palate and uvula takes a slightly different course than that of the regions of the secondary palate which give rise to the hard palate [5]. The soft palate and uvula develop from two separate masses found at the most posterior portions of the secondary palatine shelves. Unlike the fusion mechanism which is in place along much of the length of the palatine shelves, the consolidation of these two separate masses is brought about by a selective proliferation of mesenchymal cells located deep in the valley between the masses. As that proliferation, called merging, continues the valley between the two distal shelf masses is obliterated, which results in a smoothening of the contour of the soft palate and uvula. Failure of the merging process in soft palate and uvula development can result in complete or partial clefts of the soft palate and uvula.

Clefts of the palate, with or without clefts of the lip, are relatively common depending on the population group of the individual [23, 65]. Whereas occurrence figures for nonsyndromic cleft lips (with or without cleft palate) are about 1 in 1,000 live births, clefts of the palate (with or without cleft lip) occur in 1 in 2,500 live births, again depending on the population group of the individual, i.e., highest incidence in Asian, intermediate in white, and lowest in black populations [60]. Most clefts of the lip and palate generally are related to an interplay of genetic and environmental factors, i.e., multifactorial inheritance [21]. While animal studies have provided some insight into the molecular and cellular bases of these defects, precise etiologic explanations, especially involving teratogens in the

etiology of clefts of the human lip and palate, are still wanting. Crucial faulty chromosomes (see, for example, ref. 17) and genes linked to inherited forms of cleft lip and palate have recently been identified. This candidate gene is known as Interferon Regulatory Factor 6 [IRF6] [35]. Other candidate genes have been assigned some importance in palatal morphogenesis, including *MSX1*, *LHX8* *6p24* genes (associated with palatal shelf growth and differentiation); *TGFA*, *EGFR* and *HOXA2* (associated with elevation and depression of tongue); *TGFB3* and *PVRL1* (associated with associated with sequential fusion stages along the midpalatal seam), and *TGFA* and *EGFR* (associated with actual apoptosis, or “programmed cell death” events along the midpalatal seam).

Some clefts of the lip with or without cleft palate are seen regularly in a number of single mutant gene syndromes. Other clefts are associated with chromosomal syndromes, especially in trisomy 13. A complete cleft palate represents a maximum degree of clefting and is a birth defect in which the cleft extends from the incisive foramen region through the soft palate and uvula. The incisive foramen region is the demarcation used in distinguishing the two major groups of cleft lip and palate. Anterior cleft types include cleft lip, with or without a cleft of the alveolar region of the maxilla. A complete anterior cleft extends through the lip and alveolar region to the incisive foramen region. The pathogenesis of anterior clefts is related to a deficiency of neural crest-derived mesenchymal cells chiefly within the intermaxillary segment of the lip. The posterior cleft type of birth defect generally include clefts of the secondary palate that extend from the incisive foramen through the soft palate and uvula. The observation that the female secondary palate has a longer “developmental critical period” [6] than the male embryo by approximately 1 week offers some explanation why isolated cleft palate is more prevalent in females (66%) than males (34%). In general, the pathogenesis of posterior palatal clefts is related to abnormalities in a combination of events ranging from deficiencies in mesenchymal cell numbers to perturbations in the shelf extracellular matrices to abnormal elevation and fusion of the shelves, or lack thereof, as associated with a number of hypothesized teratogens, including excess doses of retinoic acids, glucocorticoids, and dioxins.

1.1 Summary

The understanding of the natural history, clinical delineation, and clinical management of birth defects involving the face, lip, and palate has progressed significantly over the last 20 years and continues to do as we move further into the 21st century. Although hu-

man craniofacial morphogenesis is clearly the culmination of a very complex series of diverse and overlapping developmental events, all of these events can be categorized into four fundamental processes which span mammalian development and are evident in the earliest beginnings of the face and palate – normal and abnormal: (1) cell *differentiation* – the process through which the myriad of “building block” cell types invoked in facial morphogenesis are generated from the single-celled zygote; (2) *morphogenesis* – the process or set of processes through which the complex form of the face and its constituent cells, tissues, and organs will emerge in a timely fashion along patternable individual and population lines; (3) *growth* – the collective results of differentiation and morphogenesis; and (4) *dysmorphogenesis and abnormal growth* – this is the most exciting of the challenges we face today as we strive to understand how environmental influences interact with and cause changes in the expression of the genetic factors governing the behavior of those cells which will give rise to the entire human body, and especially the face and palatal regions. The treatment of defective genes is very much a part of the current clinical agenda dealing with craniofacial defects. The basic scientist, the dysmorphologist, the clinician, and, importantly, those with natural or acquired craniofacial defects have gained significant advantage from the critical use of available information coming from classical and experimental studies of human morphogenesis. These advantages will continue to increase as laboratory scientists and clinician scholars move rapidly together into the world of molecular and gene biology. These approaches should and will increase our knowledge base on the patterns and underlying causes of normal and abnormal craniofacial morphogenesis – and our patients will be all the better for it. However, most researchers and treatment providers well realize that the practical transfer of new biological information on normal and abnormal development flowing from the laboratory bench to the clinical bedside may be neither easy nor timely to achieve in the effective treatment and management of craniofacial abnormalities.

Epilogue. *“And the end of all of our exploring will be to arrive where we started and know the place for the first time.”* T.S. Eliot, 1918

References

The following references have been selected from an ever-growing literature in developmental biology and craniofacial biology in particular. Perusal of any one or several of these references should lead the reader to further readings which bear on the subject of the biology of face, lip and palate development – normal and

abnormal. References noted with an (*) are texts containing illustrations that elucidate the information contained in this chapter.

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2

Prenatal Diagnosis of Oral Clefts

JORGE L. GOMEZ

With improvements in the resolution of ultrasound equipment and technique, craniofacial malformations of the fetus are now identifiable. Oral clefts are the most common facial malformation. The incidence of oral cleft has been estimated to be about 1 per 1,000 live births [1]. The antenatal diagnosis of facial clefts allows for adequate counseling and planning for prenatal care and delivery.

The first antenatal ultrasound diagnosis of cleft lip and palate was reported in 1981 [2]. Two cases were reported in the third trimester. Recently, there have been reports of antenatal diagnosis as early as 12 weeks of gestation by transvaginal ultrasound [3]. Figure 2.1 shows a transvaginal ultrasound depicting a cleft lip at 13 weeks of gestation.

The technique for early diagnosis of cleft lip and palate has been established [4]. Two planes of the fetal face are obtained. In the frontal plane, disruption of the normal midfacial architecture, with absence of the maxillary ridge normally seen slightly anterior and inferior to the orbits can be demonstrated. Also, broadening of the nasal cavity is noted. In the coronal

plane, a soft tissue mass projecting anteriorly from the midline nasal septum below the nose is seen. Once an oral cleft has been diagnosed by ultrasound, a complete anatomical survey is warranted since there have been approximately 350 syndromes, including chromosomal abnormalities, associated with facial clefting [5].

Nyberg et al. suggested an ultrasound classification for oral clefts which describes five types [6]. Type 1 includes an isolated cleft lip without palate (Fig. 2.2). Type 2 includes unilateral cleft lip and palate. Type 3 is bilateral cleft lip and palate (Fig. 2.3). Type 4 is the median cleft. Type 5 refers to clefts associated with amniotic bands or limb-body-wall complex.

The detection rate for prenatal diagnosis of oral clefts is dependent on factors such as the experience of the operator, indications for the studies, i.e., risk factors, and gestational age at the time of the study. In addition, in some prenatal ultrasound laboratories, imaging of the fetal face is done routinely, whereas in other laboratories this is not done. Examination of the fetal face is not currently included in the guidelines



Fig. 2.1. Transvaginal ultrasound of cleft lip

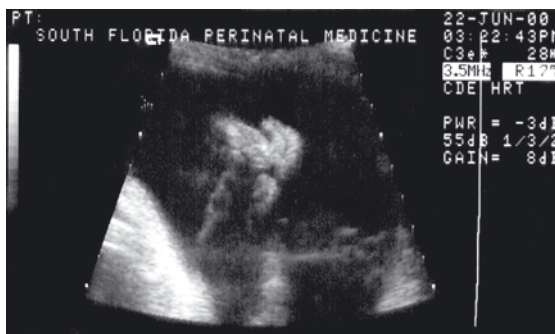


Fig. 2.2. Unilateral cleft lip



Fig. 2.3. Bilateral cleft lip and palate

for performance of the antepartum obstetrical ultrasonographic evaluation published by the American Institute of Ultrasound in Medicine [7].

Clementi et al. reported the experience in the detection of oral cleft in 20 European countries [1]. In most of the participating countries, evaluation of the fetal face was done routinely. Out of 709,027 births, there were 751 facial clefts (1 per 1000); 553 were cleft lips with or without palate and 198 were isolated cleft palate. The detection rate was 27% for cleft lip/palate and 7% for cleft palate. Fetal deaths occurred in 4% of the cases. On the other hand, Bronshtein et al. reported their experience on early detection by transvaginal ultrasound [8]. The population included 14,988 transvaginal ultrasounds performed during the 12th to the 16th weeks of gestation; 25% of the population was low risk for oral clefts and 75% had risk factors. The incidence of oral clefts was 0.8 per 1,000 (12 cases) and the detection rate was 92% (11/12).

Even in experienced hands, ultrasound has its limitations in correctly identifying the involvement of the palate. Cash and Coleman studied the detection rate in low risk population in the United Kingdom, where the fetal face is routinely evaluated [9]. There were 30 cases of oral cleft in a population of 23,577 (1.3 per 1,000). The detection rate for cleft lip and palate was 93% (13/14), 22% (2/9) for isolated cleft palate, and 67% (2/3) for isolated cleft lip. There were no cases of false positive diagnosis. The overall detection rate was 65% and the correct diagnosis was made in 71% of cases.

There is also the concern about false positive diagnosis of oral clefts. Hafner et al. performed prenatal ultrasound to detect malformations of the fetal face in 5,407 women [10]. There were 11 facial clefts identified, 8/11 (72%) were detected prenatally. However, there were also 8 false positive cases for a 99.8% specificity. It was interesting in this study that there was a

100% detection rate when the oral cleft was associated with other fetal anomalies, but 50% when it was isolated.

Gestational age at the time of the ultrasound has implications in the ability to detect oral clefts. Robinson et al. reported their experience in Brigham and Women's Hospital [11]. There were 56 confirmed cases of cleft lip in a period of 10 years. The detection rate was 57% before 20 weeks of gestation versus 80% when the ultrasound was done after 20 weeks. Therefore, in patients at increased risk for oral clefts and with a normal early ultrasound should have a repeated study after 20 weeks of gestation.

Three-dimensional ultrasound seems to aid in the correct diagnosis of oral clefts. Chen et al. evaluated 21 fetuses with confirmed oral clefts scanned between 20 to 34 weeks of gestation [12]. The accuracy (true positive + true negative) of 3D ultrasound in the diagnosis of oral clefts was 100% versus 29% for 2D ultrasound. In another study by Chmait et al., they found a 100% accuracy of 3D ultrasound versus 91% for 2D ultrasound [13]. However, the same study found a decreased specificity for cleft palate with 3D ultrasound (83%) versus 2D ultrasound (92%).

Neonates with cleft lip/palate usually do not have associated malformations; however, the incidence of severe structural anomalies, mainly central nervous system and cardiac, detected by prenatal ultrasound in association with oral clefts is increased; this is due to a high incidence of fetal demise and elective termination of pregnancy. Saltzman et al. found that 10 out of 12 cases (83%) of fetuses with prenatally diagnosed oral clefts had other structural anomalies, and 4 out of the 10 (40%) had an autosomal trisomy [14]. Benacerraf et al. described the outcome of 32 fetuses with prenatally diagnosed oral clefts [15]. Out of 32 fetus diagnosed over a 3 1/2 year period as having cleft lip/palate by ultrasonographic examination, 53% (17/32) had other sonographically detected structural abnormalities; 35% of these (6/17) had aneuploidy (five had trisomy 13 and one trisomy 18). Five pregnancies were electively terminated. Eight died in utero or in the neonatal period. Of the 15 fetuses that had no other abnormalities detected (47%), four electively terminated (27%), one miscarried, and one had vertebral abnormalities and died due to pulmonic stenosis. Nine survived (60%) and underwent successful correction of their cleft lip/palate. Nicolaides et al. found that all the fetuses diagnosed with oral clefts and chromosomal abnormalities had associated structural anomalies [16].

Nyberg et al. reported on the correlation of type of cleft with chromosomal abnormalities and associated structural anomalies [6]. They found a risk of 0% for chromosomal abnormalities and 20% for associated structural malformations in Type 1 (unilateral cleft

lip); 20% and 47% respectively for Type 2 (unilateral cleft lip and palate); 30% and 55% respectively for Type 3 (bilateral cleft lip and palate); 52% and 100% respectively for Type 4 (median cleft). There was only one fetus with a chromosomal abnormality and isolated oral cleft. Berge et al. found that the risk for chromosomal abnormalities, other structural fetal anomalies, and fetal death was 0%, 0%, and 0% respectively for Type 1; 32%, 48%, and 48% respectively for Type 2; 50%, 72%, and 65% respectively for Type 3; 82%, 100%, and 100% for Type 4. All fetuses with isolated cleft palate had abnormal chromosomes, other malformations, and died in utero or in the neonatal period.

Prenatal diagnosis of oral cleft is important for those parents at increased risk for recurrence. Ultrasound laboratories that routinely perform evaluation of the fetal face have to be familiar with the limitations of the diagnosis, i.e., accuracy of the diagnosis and false positives results. Also, these laboratories should be prepared to offer adequate counseling and referral to experts in the treatment and correction of oral clefts. Amniocentesis for isolated oral clefts remains unresolved with incidence of aneuploidy reported to be as low as 0% to as high as 5%. Proponents of routine visualization of the fetal face argue that in the future when fetal surgery becomes safer for both mother and fetus, fetuses with oral cleft may benefit from early in utero intervention.

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3

The Value of Longitudinal Facial and Dental Casts Records in Clinical Research and Treatment Analysis

SAMUEL BERKOWITZ

After 40 years of treating children with various types of clefts, this author has concluded that the success or failure of a surgical procedure depends on the degree of palatal cleft defect at the time of surgery and the resulting facial growth pattern, as well as the surgical skills and the surgical procedure utilized. This conclusion will not be new to the experienced orthodontist, who in all probability recognizes that the progress recorded in treatment depends, for the most part, on the skeletal and facial growth patterns inherent in the patient, and the interaction of surgery with facial and palatal growth.

3.1 Serial Cephaloradiographs and Casts of the Maxillary and Mandibular Dentition and Occlusion

To properly assess the results of treatment, there is a fundamental need for serial casts, lateral cephalometric films, and photographs in individual case reports.

Pruzansky [1, 2] often stated that it is unfortunate that plastic surgeons' training in the realm of clefts and their variations tends to be totally inadequate, because their first encounters with patients usually occur in the clinic or operating room. Furthermore, there is seldom recourse to anatomical specimens to better appreciate the nature of the cleft deformity. The trainee is dependent on the empirical experience of his preceptor for knowledge of the natural history of the defect and long-term response to therapy. In most cases, other than before and after facial photographs, there are no objective records to determine why the outcome was a success or failure.

The collected serial data to be shown in this text will provide the clinician in training with an overview of the variations that can be encountered in each cleft type, the significance of genotype differences that influence growth and response to surgery, and the natural history of each cleft entity.

Over the years, certain cephalometric measurements have become standardized and have been applied to selected population samples to develop statistical means or averages. In the treatment of cleft lip and/or palate, this approach has provided useful data in studying morphologic growth changes in the head, evaluating dentofacial abnormalities, and assessing responses to surgical and orthodontic treatment. The data have been particularly useful in determining the timing and type of procedure selected to treat individual problems. The measurements and analyses utilized are primarily profile-oriented and reveal both anteroposterior and vertical relationships of the various parts of the dentofacial complex.

To assess changes during the course of general growth and treatment, head radiographs of the same individual taken at separate times are traced, and the tracings superimposed to ascertain the changes that have occurred. A common method is to register the two tracings at the point sella with the sella-nasion lines superimposed (Fig. 3.2a, b). This method provides a gross overview of changes in the dentofacial complex and in soft tissue, but is useful only in evaluating what has already occurred. In this text we also use the Coben [3] superimposition procedure (Basion Horizontal) because it more accurately reflects actual craniofacial growth direction (Figs. 3.1d, 3.2c).

The use of "landmark," or baseline, images associated with the basicranium to show the composite results of facial growth can provide meaningful information, because it is the enlargement of the face relative to the cranial base that is being evaluated. In the child, further growth changes in the anterior part of the cranial base slow considerably at about 5–6 years of age, whereas facial growth continues actively through adolescence or beyond. Comparing the relative growth between these two regions, rather than simply focusing on a single fixed point, provides clinically useful information when cephalometrically evaluated.

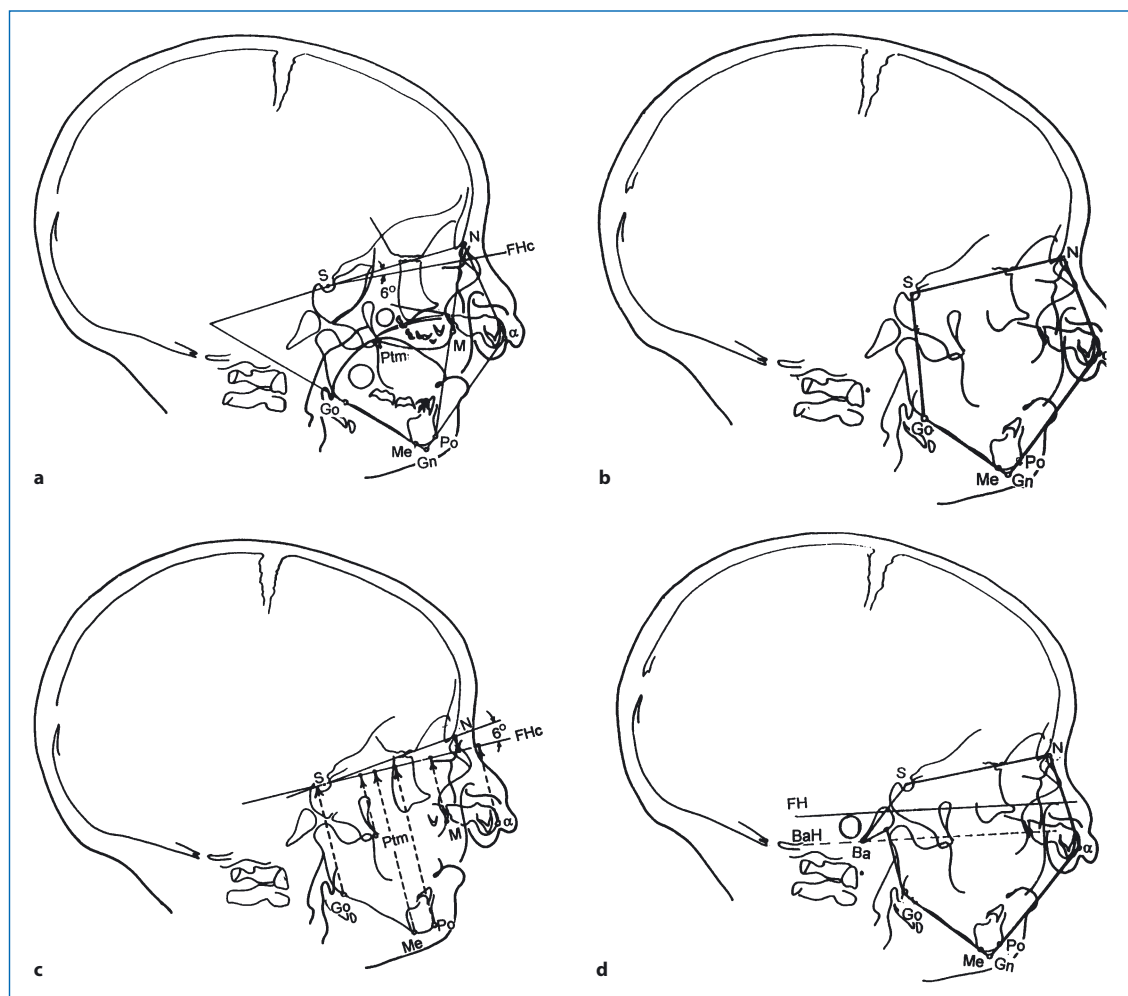


Fig. 3.1 a-d. Various methods used to demonstrate facial changes using lateral cephalometrics. **a** Facial angles. These are just a few of the angles which describe changes in the skeletal profile. There are many more angles and linear measurements which can be used to relate the maxilla to the mandible and both jaws to the cranial base. **b** Facial polygon. This is a graphic method used to describe the boundaries of the skeletal face. (Pogonion constructed, Po', is the same point as Gnathion.) Facial growth changes can be shown by superimposing each succeeding polygon on the anterior cranial base (SN) and registering on Sella turcica (S). **c** Projecting facial landmarks to a

constructed Frankfort horizontal line which is arbitrarily drawn 6° off the SN line. This angle can vary with steepness of the anterior cranial base. This graphic method will show the relative contribution of various structures within the maxilla and the mandible to the profile. **d** Basion horizontal created facial polygon (S.E. Coben, 3). This method of superimposing tracings graphically reflects his overall concept of fixed growth. A plane at the level of the anterior border of foramen magnum (Basion) parallel to Frankfort horizontal where Basion is the point of reference for the analyses of craniofacial growth

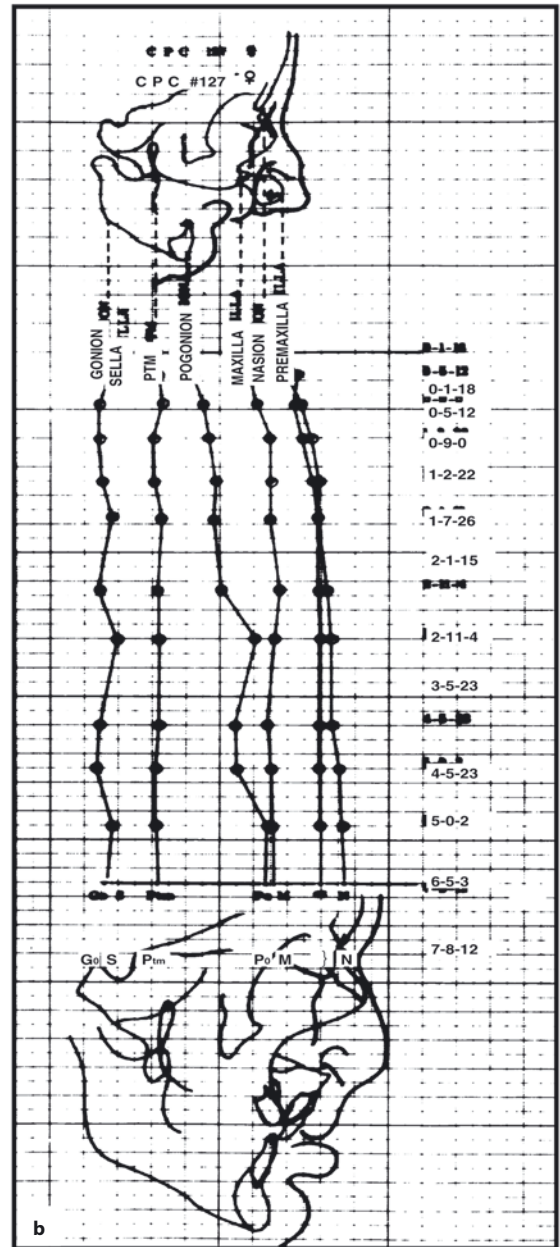
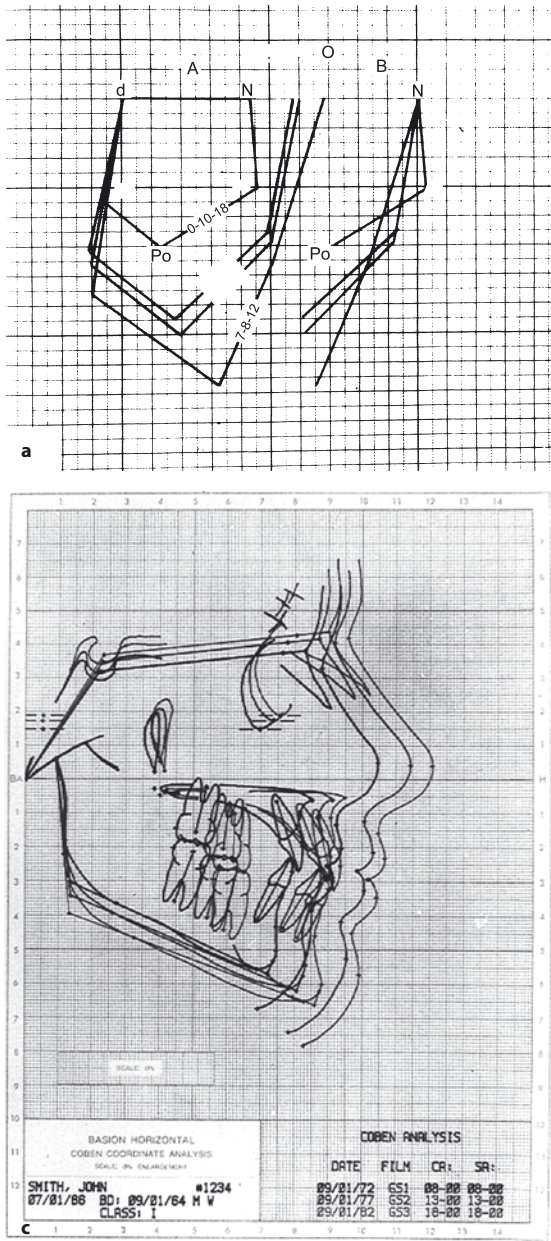


Fig. 3.2. a Case CP #127 (CPCLP). Superimposed facial polygons from 1 month, 18 days of age to 7 years, 8 months and 12 days of age. A result of the mandible's downward growth increments exceeding its horizontal growth increments, this is an example of "poor" facial growth in that the profile fails to flatten as the mandible remains retrognathic. Note that in this and the following illustration the forward projection of the premaxilla does not increase after 1 year, 2 months and 22 days. **b** Case CP #127. Projecting facial landmarks to a constructed Frankfort horizontal 6° off the SN line. Although each of the skeletal struc-

tures except for the mandible has increased in size, the relative position of midfacial structures to the anterior cranial base has remained relatively stable. **c** Case CP #127. Basion Horizontal coordinate computer craniofacial serial tracings at ages 8, 13, and 18 years. Tracings are registered at Basion and oriented in Frankfort horizontal. Serial tracings maintain a constant S-N/FH relationship. S-N and FH planes are parallel. Tracings depict Coben's growth philosophy, which states that craniofacial growth is reflected away from the foramen magnum (Basion) and the vertebral column (Reprinted from [3])

3.2 The Beginning of Longitudinal Cleft Palate Research Studies

Two major research problems were common in cleft palate surgical studies prior to the 1950s. Pruzansky [1, 2] commented on the surgeon's tendency to group all types of clefts together in research and clinical treatment. He also stated that surgeons were limited in their study of pathologic anatomy of clefts due to the unavailability of serial dental casts, cephaloradiographs, and photographic records.

The need for clinical records was apparent to many researchers, and within a decade, many retrospective clinical data sets were developed. These data sets spawned many investigators to determine the long-term influences of surgical and neonatal maxillary orthopedic procedures on palatal and facial growth and development. As a result of these early studies, useful diagnostic and prognostic information was obtained that provided a rationale for the management of individual cleft cases. These clinical records offered an accurate means for measuring and recording individual variation and for plotting the progress of each case in terms of growth and response to various treatments. As a result of these findings, the quality of care improved, resulting in more aesthetic and functional outcomes. Proper documentation, using objective records and individual treatment outcomes, has extended to many more modifications where it is possible to perform multicenter retrospective studies.

3.3 Research Methods [4, 5]

3.3.1 Retrospective Studies

In a retrospective study, the nature of the study group must be delineated precisely. Definite criteria should be established so that there is no ambiguity about types of cases and stages of growth development to be included in, or excluded from, the study. The choice of the case and control groups should be guided by concerns of validity. The advantages of retrospective studies are that they can be conducted relatively rapidly because the records of patients whose treatment is already complete can be used. The investigator is protected against the circumstance of "subject dropout" during the course of treatment, and they are relatively economical.

3.3.2 Prospective Studies

The advantages and disadvantages of prospective studies are in essence the inverse of those of retrospective studies. Provided that ethically and logisti-

cally satisfactory plans for random assignment to treatment can be developed, prospective trials afford an opportunity to control selection bias and to define and control the records acquisition process.

The main disadvantages of prospective trials are that they are expensive and a great deal of time must inevitably elapse between project initiation and the point at which data on most of the main outcome variables become available for analysis.

Multicenter comparisons of surgical-orthodontic treatment outcomes are an efficient way of testing the effectiveness of various treatment philosophies and surgical techniques. Differences among surgeons, variances in performance by the same surgeon over the years, and differences in techniques are difficult to identify and compare in isolation. However, in multicenter clinical studies, differences in clinical procedures among operators can, within defined limits, be compared and evaluated successfully without arousing criticism.

3.3.3 Clinical Trials

A clinical trial may be defined as a carefully designed, prospective study that attempts to answer a precisely defined set of questions with respect to the effects of a particular treatment. A clinical trial is a major undertaking which requires considerable money, personnel, facilities, time, and effort.

The simplest design for a clinical trial involves randomization between two different surgical treatment regimens to answer one specific question, for example, which of two surgical procedures is the most beneficial. To add a larger number of surgical procedures makes the trials more difficult to manage.

There are two reasons for not using a randomized clinical trial (RCT) method for surgical evaluation of cleft closure procedures whether done as a multicenter or single center trial. The first is the need for the surgeon to disregard the unique nature of the individual cleft defect and perform a standard surgical treatment being tested, the presumption being that clefts of all sizes and shapes will react the same way to the same surgical procedure. The second reason concerns the ethical questions involving the sequencing of surgical procedures and the use of the surgeon's skills.

3.3.3.1 Randomization of Surgical Procedures

In proposed multicenter RCT, it is expected that each surgeon will randomly utilize various surgical procedures sequentially for each type of cleft to determine the relative differences in outcome between procedures.

With the present restraints on certain types of human research, Human Subject Research Review Committees in most settings would be reluctant to permit the use of various elective surgical procedures in a research setting if there is a possibility that a surgical procedure might lead to facial disfigurement. Most surgeons would reject participating in a study employing a particular procedure they already have used and found to be inadequate. Many surgeons see the choice and timing of cleft surgical procedures as varying with the geometric characteristics of the palatal defect; therefore, the concept of randomization cannot be considered as an alternative to what they are already doing. The factor of surgical skill in a randomized trial must be considered as a variable in determining the effectiveness of a procedure. Can all participating surgeons be equally skilled in all procedures?

3.3.3.2 The Ethics of Surgical Retrospective Clinical Trials (RCT)

It is impossible to disassociate scientific from ethical considerations when dealing with cleft palate research [6–9]. Different research protocols and evaluation methods carry different ethical problems, the more so when life or death issues are not being considered.

3.3.3.3 Informed Consent

When a patient is deemed appropriate for a particular clinical trial, a first step is often to obtain informed consent. This is a legal requirement in the USA, but not in all countries. In some European countries each participating hospital decides on whether and how to handle informed consent.

Informed consent is a social construct based on ethical guidelines and supported by legal precedents. In order for consent to be legally valid, it must be obtained voluntarily from a mentally competent person of legal age.

The greater the seriousness of the potential injury, even if the risk is minimal, the greater the obligation to inform the patient (or parent). The greater the chance of a risk occurring, even if the injury would be minimal, the greater the obligation to inform the patient (parent). The more elective the proposed treatment, the more serious injury will be perceived.

Sheldon Baumrind [10], summarizing the role of clinical research in orthodontics which is also applicable to cleft palate research, states:

Cogent arguments can be made concerning the ethics of conducting structured clinical experiments in the kinds of long-term therapeutic situations which

interest orthodontists. One telling argument is that since therapists have an absolute and transcendent obligation as professionals to deliver for each patient the treatment which they believe best for that patient, no subject can ethically be randomized to one of two possible treatments unless there is true uncertainty as to which of the two treatments is in the patient's best interest. For the same reason any experimental design that asks a clinician to treat a patient against the clinician's own professional bias is inappropriate at best. And even if ethical reservations could be overcome, it would clearly be of only minimal scientific value to accumulate data on how patients fare under treatments not considered optimal at the time they are delivered.

Baumrind [10] concludes:

Except in special and very limited circumstances, clinical studies in orthodontics cannot and should not be expected to reveal categorically which of two or more treatments is better in a global sense. They can and should be expected to supply valid and reliable information about the mean effects of different treatments. But more important, they should supply information about the usual individual variability of human growth, development, and response to therapeutic intervention.

Retrospective studies have permitted clinical investigators to evaluate the palatal and facial growth and development responses within a particular cleft type according to the type and timing of the surgical procedures employed. Such studies have shown that the degree of palatal scarring is directly related to the areas of denuded bone resulting from the displacement of the palates mucoperiosteum during cleft closure.

Roentgencephalometry has aided in the elucidation of the nature of the craniofacial malformation associated with facial clefts as it affects the mandible, maxilla, orbits, nasopharyngeal area, and the base of the skull and cervical vertebrae. Moreover, current studies on the variable growth and involution of tonsils and adenoids have raised a number of questions of interest to immunologists.

3.4 The Need for Geometric and Quantitative Analysis of Cleft Palate Casts

Treatment planning in cleft lip and palate habilitation is contingent upon understanding the natural history of the palatal cleft defect and the face in which it exists. Longitudinal dental cast studies ultimately helped explain many cause and effect relationships which existed between palatal surgery and subsequent facial development. However, there still remains an important need for our understanding of palatal

development, which will further refine and improve rehabilitative procedures. The purpose is to consider previously posed questions in light of newer biostereometric techniques. Specially, the following questions have been asked:

1. Are the palatal shelves intrinsically deficient, adequate, or excessive in mass?
2. To what extent does the geometric relationship of the palatal shelves in one cleft compare with that of another in the same type of cleft? With other types of clefts? With normal palates?
3. To what extent are the palatal shelves displaced in space?
4. How are these parameters altered as a consequence of growth and surgical reconstruction? The advent of advanced biostereometric technique, 3D digital cameras with computer made it possible to analyze the size and shape of the palate in greater detail through intensive geometric survey. The data collected by these systems can be reduced to a mathematical format and subjected to analysis by high speed computers.

In accordance with these objectives, we undertook a series of phased studies utilizing stereophotogrammetric electromechanical 3D digital analysis of serial casts of infants with cleft lip and palate with the following specific aims:

1. Test the reliability of the method for selecting the proper anatomical landmarks when extrapolating data from the stereophotographs.
2. Compare and contrast 2- and 3D surface area measurements with other descriptive measurements to determine if there are significant differences in their interpretive values.
3. Perform 3D analysis of serial casts in order to describe the changing geometry of the palatal vault in mathematical terms.

4. Determine whether the descriptive analysis revealed additional information relative to the geometric changes that follow in the course of time.
5. Determine by differential analysis if a constant geometrical relationship might exist between the size and shape of the lesser to greater palatal segment in a complete unilateral cleft lip and palate.
6. The best time to determine palatal surgery based on the size of the cleft relative to the size of the body palate.

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4

Facial and Palatal Growth

SAMUEL BERKOWITZ

4.1 Maxillary and Mandibular Growth Concepts

It is not the author's intent to write a definitive treatise on facial growth and its control processes because there are better sources for such information. However, because the history of cleft palate treatment has been influenced by what clinicians think is the correct facial growth process, it behooves the author to support or refute the various facial-palatal growth concepts based on his own clinical findings.

4.1.1 Newborn Palate with a Cleft of the Lip or Palate

Is bone missing, adequate, or in excess? What is the geometric palatal relation of the palatal segments at birth? With complete clefts of the lip and palate, are the palatal segments collapsed or expanded? Can the palatal segments be stimulated to develop to a larger size by neonatal orthopedic appliances? A number of studies have attempted to determine whether the cleft palate was deficient or adequate in osteogenic tissue; unfortunately, the investigators were limited by paucity of data, lack of homogeneity in their samples, and the hazards of estimating growth from cross-sectional data.

4.1.2 Genetic Control Theory: Craniofacial Growth is Entirely Predetermined

Enlow [1] writes that, in the past, it was thought that all bones having cartilage growth plates were regulated entirely and directly by the intrinsic genetic programming within the cartilage cells. Intramembranous bone (maxillary) growth, however, was believed to have a different source of control. This type of

osteogenic process is particularly sensitive to biomechanical stresses and strains, and it responds to tensions and pressure by either bone deposition or resorption.

Tension, as traditionally believed, specifically induces bone formation. According to the traditional wisdom, when tension is placed on a bone, the bone grows locally in response. Pressure, on the other hand, if it exceeds a relatively sensitive threshold limit, specifically triggers resorption. According to this theory when muscle and overall body growth are complete, the bone attains biomechanical equilibrium, that is, the forces of the muscles are then in balance with the physical properties of the bone. This turns off osteoblastic activity, and skeletal growth ceases.

Unfortunately for traditional schools of thought, growth control in the human body is more complex than this. Moreover, it is now known that there is not a direct, one-to-one correlation between tension-deposition and pressure-resorption.

4.1.3 Functional Matrix Theory [2, 3] (Figs. 4.1, 4.2)

Enlow [1] goes on to explain that, with the development of the Functional Matrix Principle, a number of important hypotheses began to receive attention. One of these is that the "bone" does not regulate its own growth. The genetic and epigenetic determinants of skeletal developments are in the functional tissue matrix, that is, muscle, nerve, glands, teeth, neurocranial fossa, and nasal, orbital, oral, and pharyngeal cavities. This is primary while the growth of the skeletal unit is secondary. However, although the Functional Matrix Principle describes what happens during growth, it does not account for how it happens. Experiments have shown that mechanical forces are not the principal factor controlling bone growth.

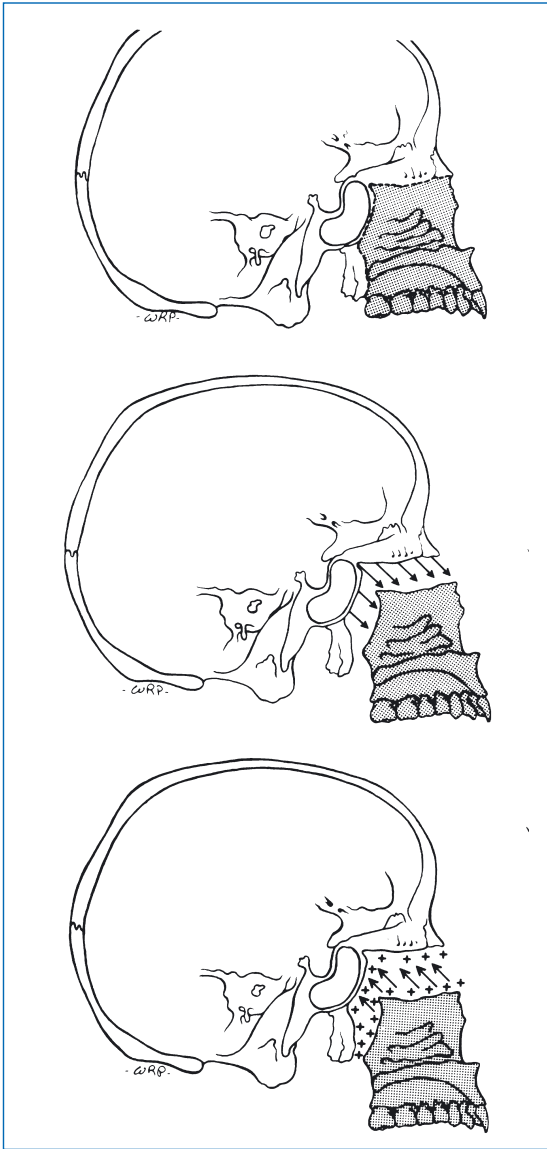


Fig. 4.1. The process of new bone deposition does not cause displacement by pushing against the articular contact surface of another bone. Rather, the bone is carried away by the expansive force of all the growing soft tissues surrounding it. As this takes place, new bone is added immediately onto the contact surface, and the two separate bones thereby remain in constant articular junction. The nasomaxillary complex, for example, is in contact with floor of the cranium (*top*). The whole maxillary region, in toto, is displaced downward and forward away from the cranium by the expansive growth of the soft tissues in the mid-facial region (*center*). This then triggers new bone growth at the various sutural contact surfaces between the nasomaxillary composite and the cranial floor (*bottom*). Displacement thus proceeds downward and forward as growth by bone deposition simultaneously takes place in an opposite upward and backward direction (i.e., toward its contact with the cranial floor). (From [1])

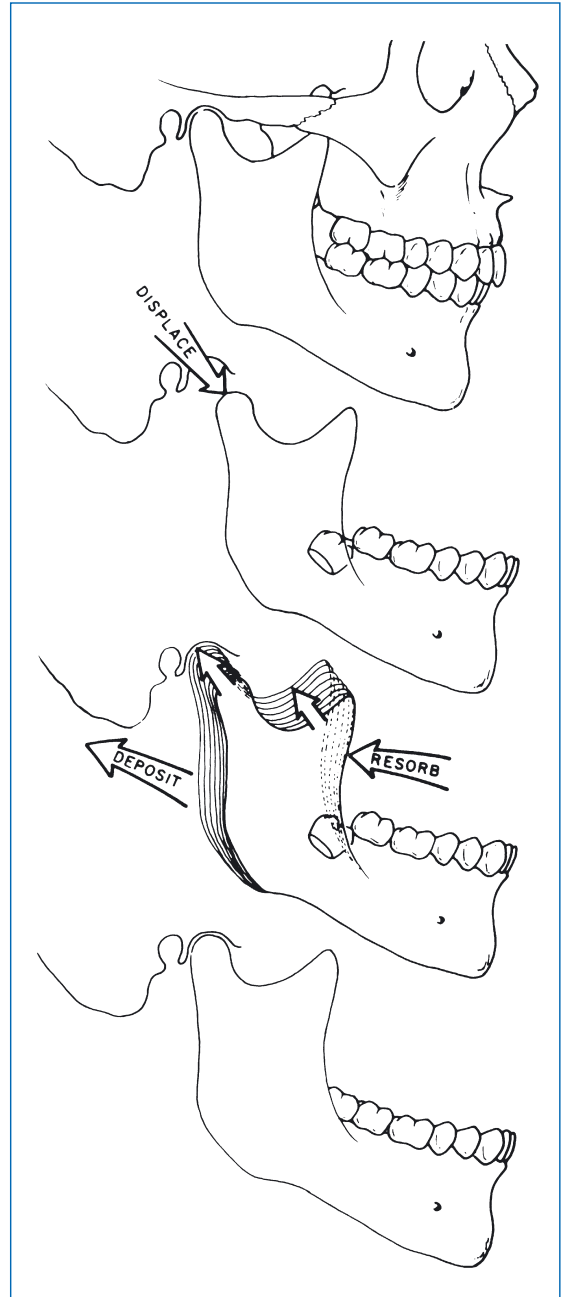


Fig. 4.2. Similarly, the whole mandible is displaced “away” from its articulation in each glenoid fossa by the growth enlargement of the composite of soft tissues in the growing face. As this occurs, the condyle and ramus grow upward and backward into the “space” created by the displacement process. Note that the ramus “remodels” as it relocates posterosuperiorly. It also becomes longer and wider to accommodate (1) the increasing mass of masticatory muscles inserted onto it, (2) the enlarged breadth of the pharyngeal space, and (3) the vertical lengthening of the nasomaxillary part of the growing face. (Reprinted with permission from [1])

Most researchers agree that a notable advance was made with the development of the Functional Matrix Principle introduced by Moss [2, 3]. It deals with what determines bone and cartilage growth in general. The concept states, in brief, that any given bone grows in response to functional relationships established by the sum of all the soft tissues operating in association with that bone. This means that the bone itself does not regulate the rate and direction of its own growth; the functional soft tissue matrix is the actual governing determinant of the skeletal growth process.

The course and extent of bone growth are secondarily dependent on the growth of pace-making soft tissues. Of course, the bone and any cartilage present are also involved in the operation of the functional matrix, because they give essential feedback information to the soft tissues. This causes the soft tissues to inhibit or accelerate the rate and amount of subsequent bone growth, depending on the status of the functional and mechanical equilibrium between the bone and its soft tissue matrix. The genetic determinants of the growth process reside wholly in the soft tissues and not in the hard part of the bone itself.

The Functional Matrix Concept is fundamental to an understanding of the overall process of bone growth control. This concept has had a great impact in the field of facial biology. The concept also comes into play as a source for the mechanical force that carries out the process of displacement. According to this now widely accepted explanation, the facial bones grow in a subordinate relationship with all the surrounding soft tissues. As the tissues continue to grow, the bones are passively (i.e., not of their own doing) carried along (displaced) with the soft tissues attached to the bones by Sharpey's fibers. Thus, for the nasomaxillary complex, the expansion of the facial muscle, the subcutaneous and submucosal connective tissues, the oral and nasal epithelia lining the spaces, the vessels, and the nerves, all combine to move the facial bones passively along with them as they grow. This continuously places each bone and all of its parts in correct anatomic positions to carry out its functions. Indeed, the functional factors are the very agents that cause the bone to develop into its definite shape and size and to occupy the location it does.

Growth control is determined by genetic influences and biomechanical forces, but the nature of the balance between them is still, at best, uncertain. No single agent is directly responsible for the master control of growth; the control process encompasses many factors. It involves a chain of regulatory links. Moreover, not all of the individual links are involved in all types of growth changes.

Enlow [1] identifies the maxillary tuberosity as being a major site of maxillary growth. It does not, however, provide for the growth of the whole maxilla,

but rather is responsible for the lengthening of the maxillary arches. The whole maxilla is displaced in an anterior direction as it grows and lengthens posteriorly. However, the nature of the force that produces this forward movement is a subject of great controversy. The idea that additions of new bone on the posterior surface of the elongating maxillary tuberosity "push" the maxilla against the adjacent pterygoid plates has been abandoned.

Bones do not by themselves have the physiological capacity to push away bones. Another theory held that bone growth at the various maxillary sutures produces a pushing apart of the bones, with a resulting thrust of the whole maxilla downward and forward. This theory has also been rejected because bone tissue is not capable of growth in a field that requires the amount of compression needed to produce a pushing type of displacement. The sutural connective tissue is not adapted to a pressure-related growth process. It is believed that the stimulus for sutural bone growth is the tension produced by the displacement of the bone. Thus, the deposition of new bone is a response to displacement rather than the force that causes it. Although the "sutural push theory" is not tenable, Enlow reports that some students of the facial growth control processes are looking anew at growth mechanizing sutures, but not in the old conceptual way.

4.1.4 Cartilage-Directed Growth: Nasal Septum Theory [4–12]

Cartilages are the leading factor. Synchondrosis, nasal septum, and mandibular condyles are actual growth centers. Sutural growth is compensatory. This theory developed from criticisms of the "sutural theory." Scott [4, 5] believes that cartilage is specifically adapted to certain pressure-related growth sites, because it is a special tissue uniquely structured to provide the capacity for growth as a result of compression. The basis for this theory is that the pressure-accommodating expansion of the cartilage in the nasal septum is the source of the physical force that displaces the maxilla anteriorly and inferiorly. This, accordingly to Scott's hypothesis, sets up fields of tension in all the maxillary sutures. The bones then, while they enlarge at their sutures in response to the tension created by the displacement process, move in relation to each other.

The nasal septum hypothesis was soon adopted by many investigators in cleft palate centers around the world and became more or less the standard explanation, replacing the "sutural theory." Clinicians involved in cleft palate treatment, such as McNeil [13–15] and Burston [16–19] and their followers [20–34] accepted Scott's thesis that cartilage and

periosteum carry an intrinsic genetic message that guides their growth. They believed that the cartilaginous centers, such as the chondrocranium, the associated synchondroses, and the nasal septum, should be viewed as the true centers of skull and facial growth. Scott [4, 5] further suggests that the nasal septum plays more than a secondary role in the downward and forward vector of facial growth.

McNeil [13, 14], following Scott's thesis, describing the embryopathogenesis of complete clefts of the lip and palate and their treatment at the neonatal period, wrote that the palatal processes, being detached from the growing nasal septum, do not receive their growth impetus and, therefore, are not only retruded within the cranium but are also deficient in osteogenic tissue. He goes still further and believes that the deficient palatal processes can be stimulated to increased size through the use of functional orthopedics.

4.1.4.1 Stimulation of Bone Growth – Is it Possible?

As McNeil saw it, pressure forces created by “functional” orthopedic appliances, which are within the limits of tolerance, will act to stimulate bone growth in an anterior direction. This force needs to be applied to particular regions and in particular directions so that it can intensify normal forces. The resulting narrowing of the cleft is due to growth of the underlying bone brought on by such stimulating appliances. Additional growth leads to a reduction in the soft palate cleft as well, thereby increasing the chance of having a long, flexible, well-functioning soft palate after surgical closure.

McNeil [14] goes on to suggest that an obturator alone is unsatisfactory because it will reduce “valuable” tongue space and lead to harmful speech habits. McNeil was correct in stressing that surgery should be reduced to a minimum compatible with sound clinical reasoning and accepted surgical principles.

Whereas McNeil states that his procedure stimulates palatal growth, thereby narrowing the cleft space, Berkowitz's [35] 3D palatal growth studies – using a sample of cases that have not had neonatal maxillary orthopedic treatment and a control sample of noncleft cases – show that growth occurs spontaneously. This is an expression of the palate's inherent growth potential, which can vary among patients. Berkowitz concluded that “catch-up growth” can occur after palatal surgery (with minimum scarring) is performed (see Chap. 16).

4.1.4.2 The Need to Prevent Collapse

McNeil [13–15] further believes that the palatal segments should be manipulated to an ideal relationship prior to lip surgery to prevent them from moving too far medially and becoming collapsed with the buccal segments in crossbite. This, he suspects, will lead to abnormal movements of the tongue and give rise to faulty respiratory, sucking, and swallowing patterns, also causing abnormal growth and development of the palatal structures.

Mestre et al. [36], studying palatal size in a cleft population that had not been operated on, report that the development of the maxilla appears to be normal in unoperated cases. They do conclude that it is the type, quality, and extent of the surgery that determines the effect on maxillary growth and that osteogenic deficiency does exist to varying degrees. Our research on serial palatal growth changes supports this conclusion that palates with clefts are highly variable in size, shape, and osteogenic deficiency.

Unfortunately, McNeil's interpretation of the effects of clefting on the various vegetative functions, and in reducing palatal growth, has not been supported by controlled objective research. The inability of the manipulated arch to remain intact after lip surgery, and not move medially into a collapsed relationship, has led many clinicians to question the accuracy of McNeil's other stated benefits such as reduction of middle ear infections.

McNeil [13–15] made other faulty observations. Among them:

1. He mistakenly believed that the orthopedic appliance will stimulate the underdeveloped cleft segment in unilateral clefts of the lip and palate (UCLP) to move forward, to make contact with the premaxillary portion of the greater segment and both palatal segments in bilateral clefts of the lip and palate (BCLP), after the lip is united. Even as early as the 1960s, many orthodontists found the opposite to be true. In UCLP, the premaxillary portion of the larger segment moves medially and backward to make contact with the lesser segment due to the action of compressive lip muscle forces. If McNeil had had the benefit of serial casts, his interpretation of clinical events would, I am confident, have been totally different.
2. McNeil's claim that the lesser segments in UCLP, and both segments in BCLP, can be stimulated to grow forward is totally erroneous. His conclusions were based on conjecture, not on objective data. The results of Berkowitz's 3D palatal growth studies [37] show marked acceleration in palatal growth during the first 2 years without orthopedic treatment, with most of the growth changes occurring at the area of the maxillary tuberosity and not

at the anterior portion of the palate except for alveolar growth associated with canine development (Fig. 4.2). Movement of the cleft palatal segment anteriorly is only possible as a result of reactive mechanical forces being applied through the use of pinned maxillary orthopedic appliances or from a protraction facial mask.

One last but significant characterization of a newborn cleft of the lip and palate needs to be refuted. McNeil states that “in BCLP lateral segments are collapsed toward the midline before birth.” However, he does not explain the dynamics that can make this possible. How can segments be collapsed if there are no inwardly directed forces from the cleft lip-cheek muscle complex, especially when the tongue fits within the cleft space and acts to move the palatal segments apart?

Enlow's [1] report on current thinking on palatal growth processes delivers McNeil's thesis a mortal blow. Enlow[1] writes that recent research has shown that pressure is detrimental to bone growth.

Bone is necessarily both a traction- and pressure-adapted kind of tissue. The periosteal membranes are constructed to function in a field of tension (as by the pull of a muscle). Covering membranes are quite sensitive to direct compression because any undue amount causes vascular occlusion and interference with osteoblastic formation of new bone. Osteoclasts, conversely, function to “relieve” the degree of pressure by removing bone. Bone is pressure sensitive and high-level pressure induces resorption.

Moss et al. [38], responding to the role of nasal septal cartilage in mid-facial growth as put forth by Scott [4, 12], states that Scott's hypothesis is based on the following assumptions: (1) that in the fetal skull, the original nasal capsule and its derivatives are cartilaginous; (2) that all cranial cartilaginous tissues (septal, condylar, or in synchondroses) are primary growth centers, by virtue of the undoubted ability of all cartilaginous tissues to undergo interstitial expansive growth; and (3) that following the prenatal appearance of the intramembranous vomer (and of the several endochondral ossification centers of the ethmoid sinuses and the turbinates) the remaining unossified portions of the cartilaginous nasal capsule continue to be capable of such interstitial expansion. Moss further suggests that the nasal septal cartilage grows as a secondary, compensatory response to the primary growth of related oro-facial matrices and that mid-facial skeletal growth is not dependent on any prior, or primary, growth “impetus” of the nasal septal cartilages.

In Scott's hypothesis, it is assumed that cartilaginous interstitial growth is the major source of the expansive force that “pushes” on the subjacent mid-fa-

cial skeletal structures, causing both vertical and anteroposterior growth. Moss believes that it has been demonstrated repeatedly that growth in size and shape, as well as the changes in spatial position, of all skeletal units is always secondary to primary changes in their functional matrices. This secondary skeletal unit growth comes about in the following manner. All cranial bones and cartilages originate and grow within soft tissue capsules. The splanchnocranial skeleton exists within an oro-facial capsule. The primary growth of the enclosed oro-facial matrices causes the oro-facial capsule to expand responsively. Because the splanchnocranial bones are within this capsule, they are passively translated in space within their expanding capsule. As a result of such spatial displacement, the individual bones will be distracted (or separated) passively from one another.

The increments of growth observed at the sutural edges of these bones, and at the mandibular condylar cartilages, are secondary, compensatory, and mechanically obligatory responses of the skeletal units to such separative movements (i.e., the alterations of size and shape in bones and cartilages are responses to matrix growth, not the cause of it).

The nasal skeleton is characterized by a relatively great normal variation in form. The nasal capsule (and septum), from its inception, serves to protect and support the functional spaces for respiration and olfaction. In human, the olfactory spaces are fully formed at birth. Postnatal cavity growth exclusively increases the respiratory functioning space.

The growth of the upper face is, in part, a response to the functional demands for increased respiratory volume. The nasal cavity is not a space haphazardly left over after the upper facial structures complete their growth. On the contrary, the expansion of the nasal cavity is the primary morphogenetic event; and nasal capsular growth, both osseous and cartilaginous, is secondary. The application of the theory of functional cranial analysis to nasal and mid-facial skeletal growth demonstrates that the growth of each of these two areas is independent of the other and that the nasal septal cartilage plays a secondary compensatory role, rather than a primary morphogenetic one.

At present, the nasal septum theory is somewhat accepted as a reasonable explanation by a number of clinicians who favor presurgical orthopedic treatment, although it is universally realized that much more needs to be understood about facial growth processes [39]. (The use of presurgical orthopedic treatment is covered in greater detail in Chaps. 18–22.)

Clinically, there seems to be more support for the functional matrix theory than the nasal septum theory. Unfortunately, McNeil, in espousing Scott's theory to explain the “retropositioned maxillary complex rel-

ative to the mandible and osteogenically deficient palatal processes” in complete clefts of the lip and palate, did not have access to serial palatal and facial growth records to support such a view. However, Berkowitz’s [40] serial casts study of CUCLP and CB-CLP cases using the Angle occlusal classification system, which is the most reliable means of judging the geometric relationship of the maxillary to the mandibular arches within the face, showed that at 3–6 years of age, the teeth in the lateral palatal segments were in either a Class I or Class II relationship but were never in a Class III relationship.

On this basis, one can conclude that it is not the lack of a growth impetus from the nasal septum that explains the presence of a small cleft palatal segment at birth. If palatal osteogenic deficiency does exist, it can more accurately be explained in relationship to the embryopathogenesis of facial development: the failure of migrating undifferentiated mesenchymal cells from the neural crest to reach the facial processes [41, 42] (see Chap. 1).

4.1.5 Basion Horizontal Concept: The Direction of Facial Growth (Figs. 4.3–4.5) [43]

No discussion on craniofacial growth is complete without including Coben’s Basion Horizontal Concept of the direction of facial growth. Basion Horizontal is a concept based on a plane at the level of the anterior border of foramen magnum parallel to Frankfort horizontal where Basion is the point of reference for the analysis of craniofacial growth. Coben states that the growth concept which Basion Horizontal represents is that craniofacial growth is reflected away from the foramen magnum (Basion) and the vertebral column. The cranio-maxillary complex housing the maxillary dentition is translated upward and forward from Basion by growth of the cranial base. Growth of the mandible is reflected away from Basion, carrying the mandibular dentition downward and forward. The divergence of the two general vectors develops space for vertical facial growth and the eruption of the dentition.

Normal maxillo-mandibular development requires synchronization of the amount, timing, and direction of growth of the cranio-maxillary complex and of the mandible. The cranial base vector represents the upward and forward translation of the upper face by growth of the spheno-occipital synchondrosis, while growth of the spheno-ethmoidal/circumaxillary suture system and the nasal septum increases the depth and height of the upper face.

The Basion–Articulare dimension is essentially stable postnatally, indicating that the mandible main-

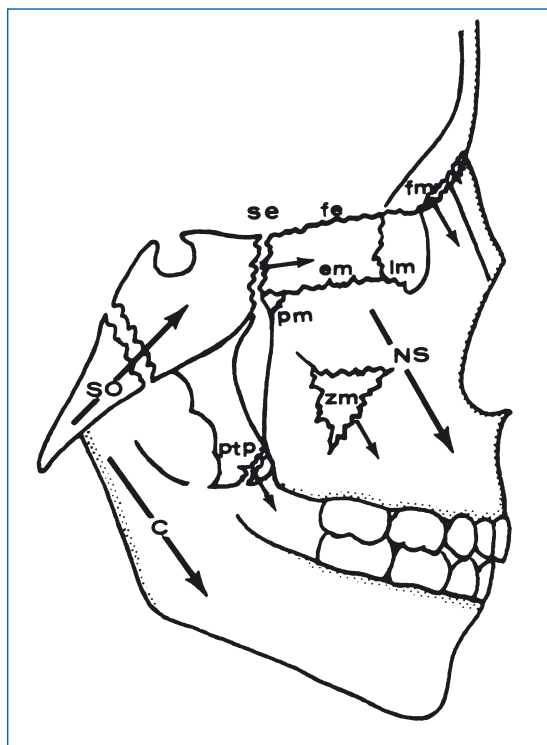


Fig. 4.3. Postnatal craniofacial growth systems to the age of 7 years (first decade). Cartilaginous growth: SO, Spheno-occipital synchondrosis; C, reflection of condylar mandibular growth; NS, nasal septum. Spheno-ethmoidal circumaxillary suture system: se, Spheno-ethmoidal; ptp, pterygopalatine; pm, palatomaxillary; fe, fronto-ethmoidal; em, ethmoidal-maxillary; lm, lacrimal-maxillary; fm, frontomaxillary; zm, zygomaticomaxillary; zt, zygomaticotemporal (not shown). Surface apposition-modeling resorption development (stippled area): minor contribution. (Reprinted from [43])

tains a constant sagittal spatial relation to the foramen magnum as the reflection of mandibular growth carries the lower teeth downward and forward, away from the cranial base.

There are two distinct phases of craniofacial growth because of a change in the system of upper facial development after the approximate age of 7 years. Before age 7, growth of the upper face is dominated by the nasal septum, the eyeballs, and the spheno-ethmoidal/circumaxillary suture system (Fig. 4.4). At this age, the growth in this suture system produces space for the eruption of the maxillary first molars. Longitudinal cephalometric findings of a continuous increase in the Sella–Frontale dimension with little increase in the thickness of the frontal bone before age 7 support the concept that bone apposition and remodeling resorption are minor factors in these early years.

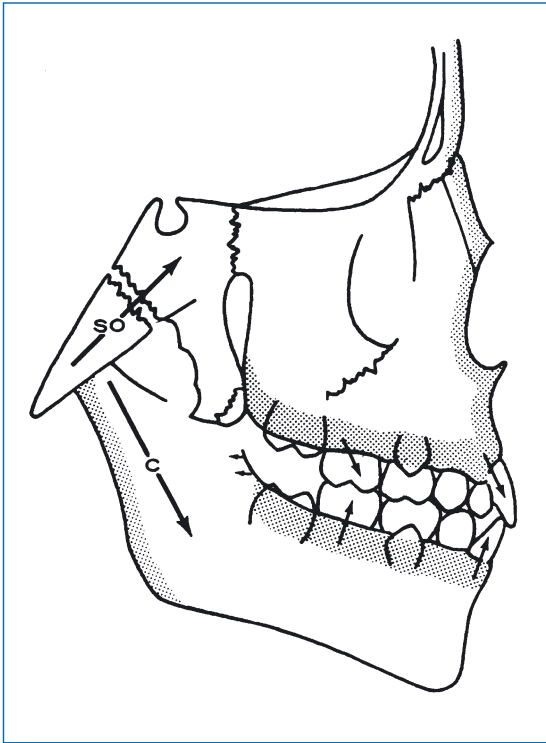


Fig. 4.4. Postnatal craniofacial growth systems from age 7 years (second decade). Cartilaginous growth: SO, Spheno-occipital synchondrosis-active through puberty; C, reflection of condylar mandibular growth – active to facial maturity; nasal septum – growth completed. Spheno-ethmoidal circumaxillary suture system: Sutural growth no longer primary system of upper facial development. Surface apposition-modeling resorption development (*stippled area*): Now major method of upper facial development and alveolar growth [43]

At about age 7, the growth system of the upper face changes with the closure of the spheno-ethmoidal suture. The Sella–Frontale dimension stabilizes, and the thickness of the frontal bone begins to increase by surface apposition and remodeling until maturity. The interpretation is that after age 7, the initial primary system of spheno-ethmoidal/circumaxillary sutural growth of the upper face is replaced by surface apposition and remodeling resorption (Fig. 4.4). It is significant that, before age 7, space for the erupting upper first molars results from growth of the spheno-ethmoidal/circumaxillary suture system. After age 7, space for the upper second and third molars is produced by maxillary alveolar apposition as the maxillary dentition erupts downward and forward. This concept was supported by Scott [12], who reasoned that the spheno-ethmoidal suture must be viewed as

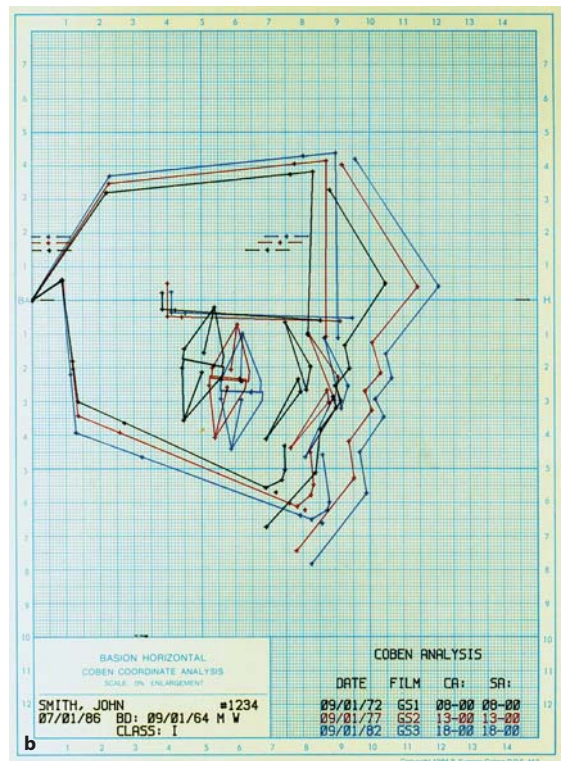
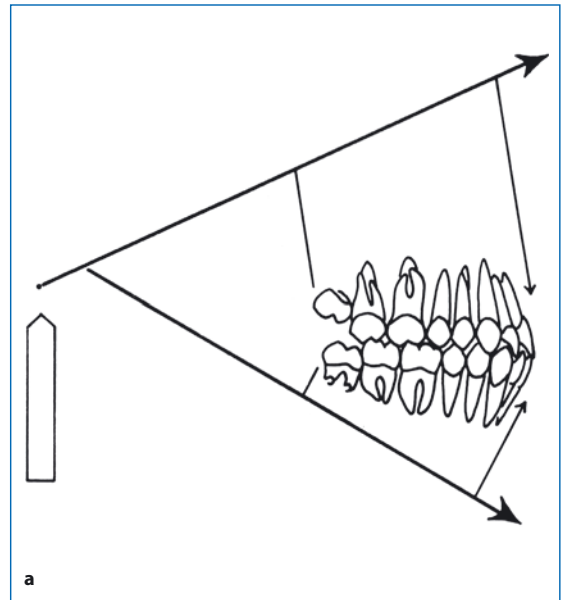


Fig. 4.5. a Basion Horizontal. General vectors of craniofacial growth. Growth of the cranial base translates the upper face and the maxillary dentition upward and forward away from the foramen magnum. Growth of the mandible translates the lower dentition downward and forward. The two diverging vectors create space for vertical facial development and tooth eruption. [43] **b** Basion Horizontal. Basion Horizontal Coordinate computer Craniofacial serial schematic line graph of Fig. 4.5a

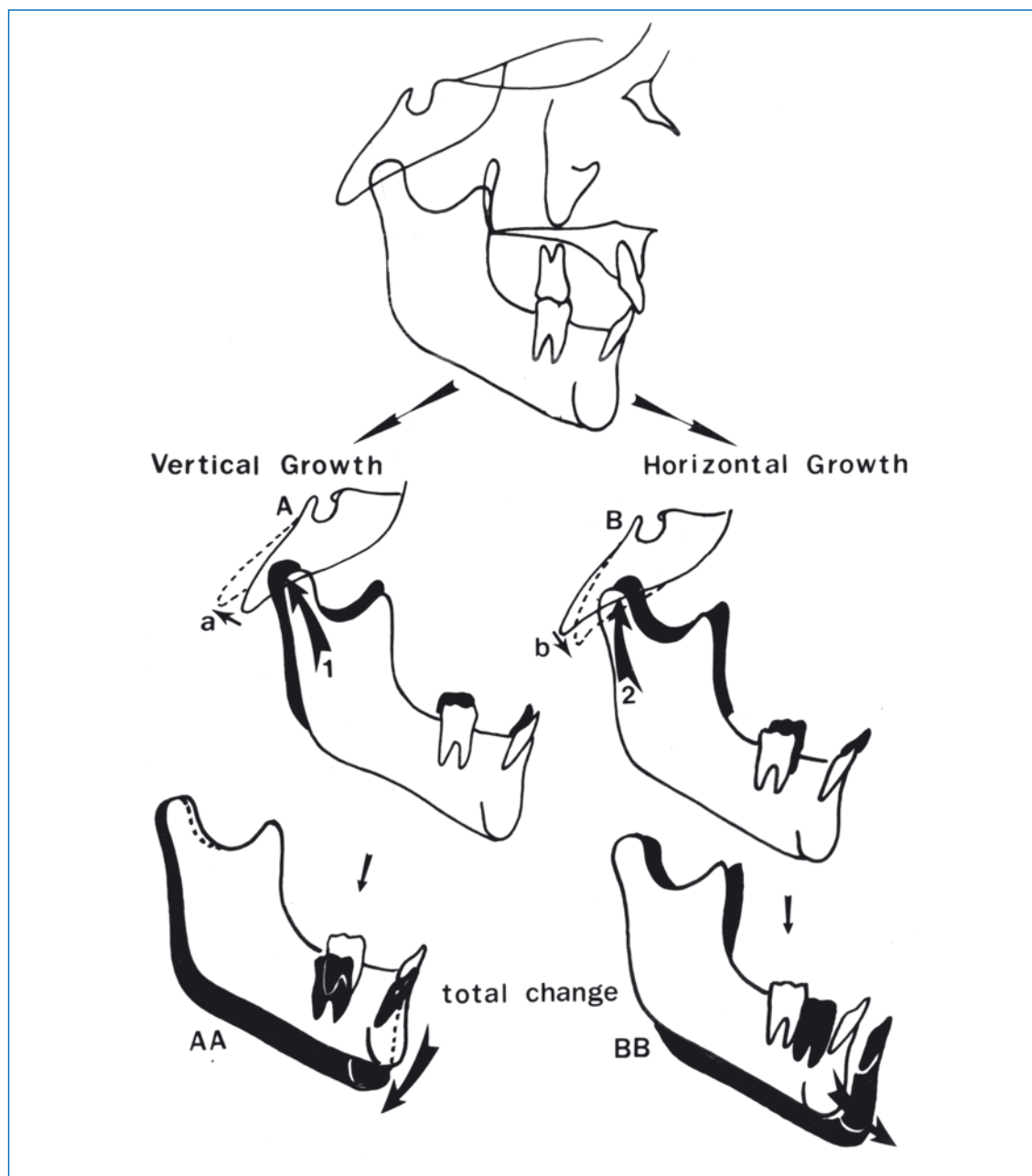


Fig. 4.6. Various growth changes that occur, the condylar head determine the direction and extend of mandibular growth

part of the major circumaxillary suture system, and that once part of the suture closes, there is no further growth in that suture system. Longitudinal cephalometric growth studies confirm this interpretation (Fig. 4.5).

4.2 Mandibular Development in Cleft Palate (Figs. 4.6, 4.7)

Recent studies have revealed a series of often subtle differences in the morphology of the mandible in persons with cleft lip and/or palate. Dahl [44] and Chierici and associates [45] found that, in persons with

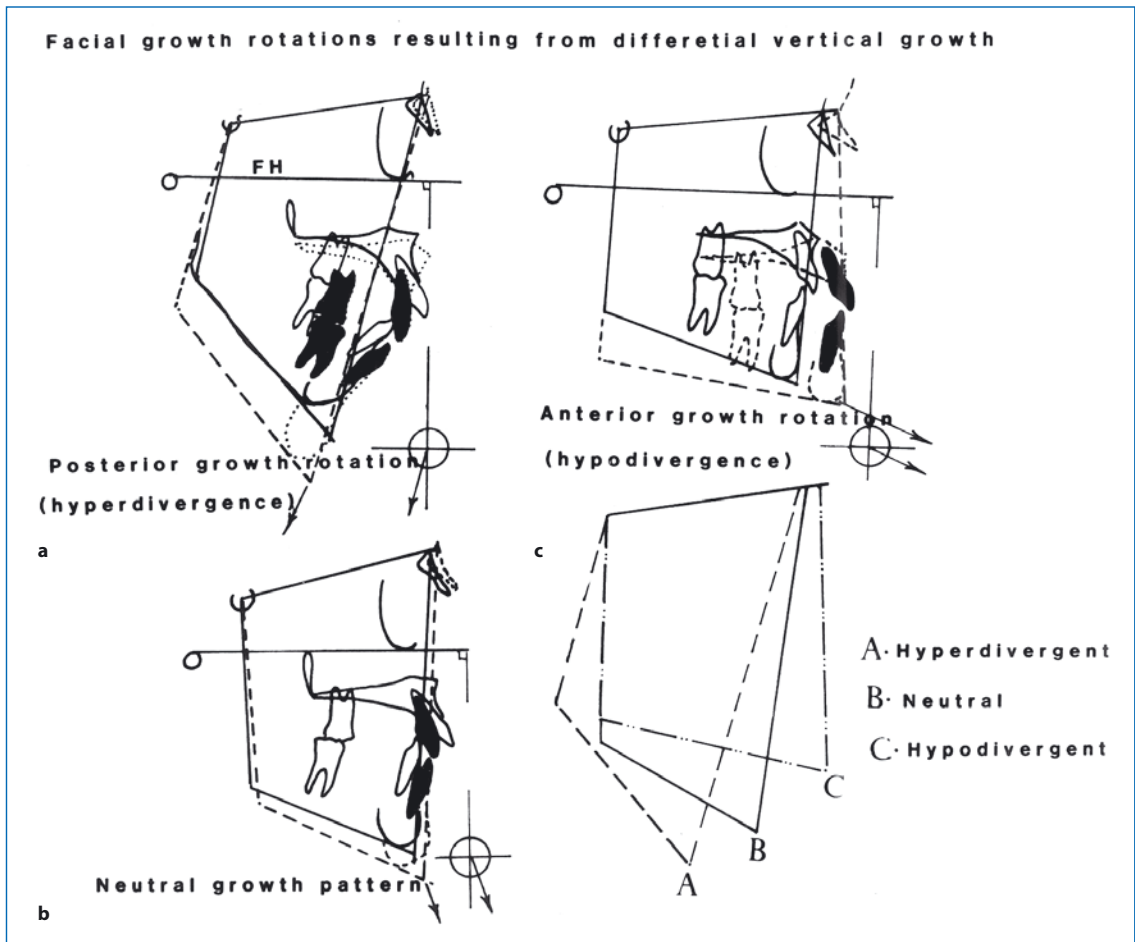


Fig. 4.7 a–c. Facial growth rotations resulting from differential vertical growth. **a** Hyperdivergent pattern with posterior growth rotation. **b** Neutral growth pattern. **c** Hypodivergent growth pattern with anterior growth rotation. *Comment: This*

series is not a true reflection of the growth of various components of the face. See Cohen's Basion Horizontal, Coordinate Craniofacial Analysis system for this (Fig. 4.5)

clefts of the hard palate only, the mandibular plane was steeper and the gonial angle more obtuse than in a normal population. Mazaheri and coauthors [46] noted that the length and width of the mandible were significantly less in persons with cleft palate only than in those with cleft lip and palate (CLP) and normal groups. Aduss [47] observed that the mandibular gonial angle in patients with unilateral CLP was more obtuse, and that the anterior cranial base appeared to be elevated. Rosenstein [48] also found the mandibles to be smaller, with steeper mandibular plane angles. Bishara [49] studied Danish children with repaired cleft palates only. In that study, and again in a later study of patients with CUCLP [50], he noted that the mandible was significantly more posterior in relation to the cranial base and that its mandibular plane was steeper than normal.

Krogman and colleagues [51] found no difference in mandibular dimensions in the BCLP population, other than a more obtuse gonial angle. They also found the temporomandibular joint to be positioned farther back, so that its effective length was less than in the normal population. Robertson and Fish [52], comparing mandibular arch dimensions, found no significant differences between normal and cleft children either at birth or at 3 years of age.

4.3 Patterns of Postnatal Growth

Based on the serial studies, three general patterns of postnatal growth have been demonstrated. In the Pierre Robin sequence, and in complete bilateral clefts of the lip and palate, most cases demonstrate substan-

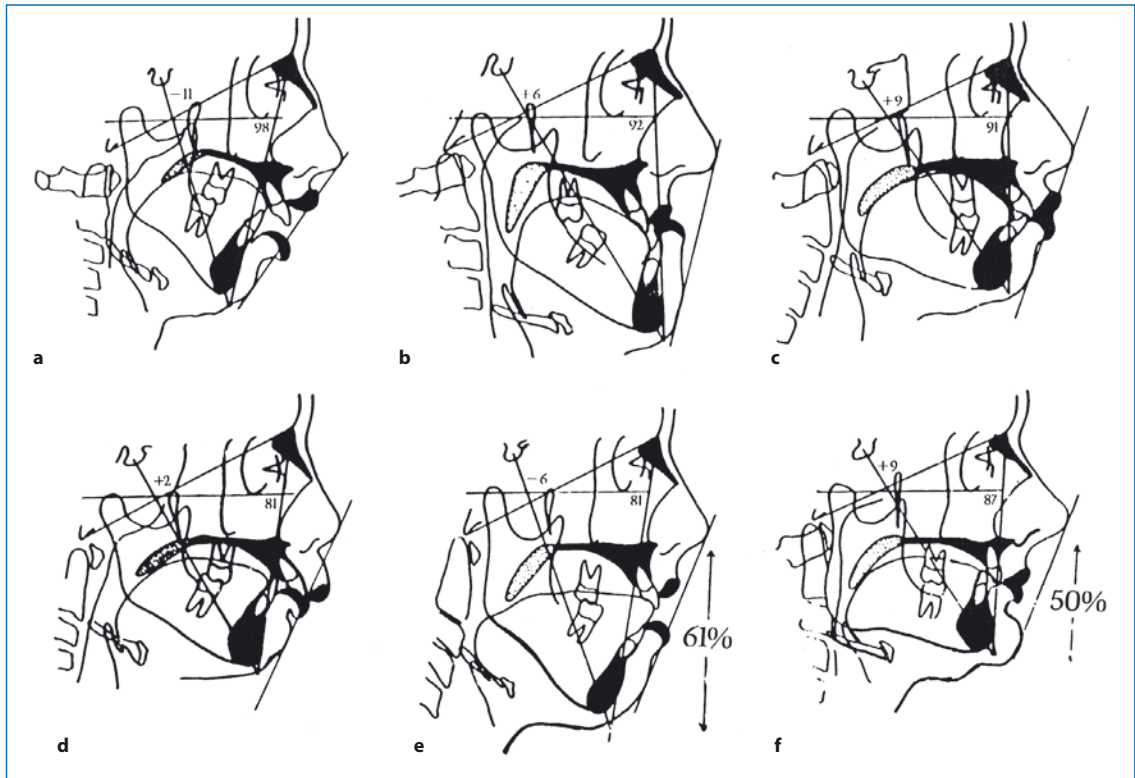


Fig. 4.8 a–f. Not all faces are the same, therefore treatment must vary according to the facial growth pattern. Various types of facial patterns. **a** Retrognathic mandible with steep mandibular plane angle. Severe overbite and overjet. Chronic mouth breather. **b** Prognathic mandible with recessive maxilla. **c** Brachyfacial type with dental protrusion. **d** Slightly retro-

gnathic type with protrusive maxillary denture and severe deep bite. **e** Long shallow face with severe tongue problem, extremely wide openbite, and an inability to close the lips. **f** Extremely closed bite with short denture height. (Courtesy of R. Ricketts. *The Biology of Occlusion and the Temporomandibular Joint in Modern Man*, 1972)

tial improvement through “catch-up” in the growth of the mandible. In the second pattern, mandibulofacial dysostosis, the pattern of growth is such that the deformity observed in infancy or early childhood is maintained throughout the growth period. The deformity of the mandible neither improves nor worsens in the course of time. The third pattern is one in which the growth process is so deranged that the severity of the deformity increases with age. This has been observed in some instances of unilateral agenesis of the mandibular ramus (e.g., hemifacial microsomia) and in the growth of the maxilla and neurocranium in some forms of premature craniofacial synostosis.

4.3.1 Bone Remodeling During Growth (Fig. 4.8)

Enlow [1] states that remodeling is a basic part of the growth process. The reason why a bone must remodel during growth is because its regional parts become moved; “drift” moves each part from one location to another as the whole bone enlarges. This calls for sequential remodeling changes in the shape and size of each region. The ramus, for example, moves progressively posteriorly by a combination of deposition and resorption. As it does so, the anterior part of the ramus becomes remodeled into a new addition for the mandibular corpus. This produces a growth elongation of the corpus. This progressive, sequential movement of component parts as a bone enlarges is termed relocation. Relocation is the basis for remodeling. The whole ramus is thus relocated posteriorly, and the posterior part of the lengthening corpus becomes relocated into the area previously occupied by the ra-

mus. Structural remodeling from what used to be part of the ramus into what then becomes a new part of the corpus takes place. The corpus grows longer as a result.

4.3.2 Maxillary Growth

The maxilla grows downward and forward from the cranial base with growth occurring at the articulations with other bones (i.e., the sutures). Björk [53] stated that during growth the maxilla is displaced in a rotational manner relative to the cranial base; however, this rotational aspect is small, which results in the downward and forward effect. Furthermore, he emphasized that there is little variation in the upper facial height between groups. Therefore, because of the small variation, it is likely that individual difference in facial form results from growth in other facial areas where there is more variation.

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5

Alternative Method Used to Correct Distorted Neonatal Cleft Arch Forms

SAMUEL BERKOWITZ

5.1 Effects of Reversing the Facial Force Diagram

The influence of soft-tissue forces on palatal form and growth has been the topic of several studies. Ritsila and coauthors [1] reported that there was “slight shortening” of the maxilla, “marked shortening” of the body of the mandible, and alterations of several mandibular angles after closure of the lip.

As perhaps an interesting footnote [1, 2], physical changes to the palate in clefts of the lip and palate in animals are very similar to the corresponding changes that are seen in humans. Bardach et al. [1, 2] studied lip pressure changes following lip repair in infants with unilateral clefts of the lip and palate. They confirmed the belief that lip repair significantly increases lip pressure when compared with a noncleft population.

Berkowitz’s [3, 4] data demonstrated that the force of the united lip against the protruding premaxilla in complete bilateral clefts of the lip and palate (CBCLP) acts first to bring about premaxillary ventroflexion. After 2–3 years there is some appearance of midfacial growth retardation to various degrees. There is strong evidence that uniting the lip does not “telescope” the premaxilla into the vomer, whereas mechanical premaxillary retraction “telescopes” the premaxilla in almost all instances (see the Chap. titled Neonatal Maxillary Orthopedics). In very rare instances it may even cause a vomer fracture.

5.2 Variations in the Palate’s Arch Form

The size and relationship of the palatal segments to each other are highly variable (see the Chap. titled The Effect of Clefting of the Lip and Palate on the Palatal Arch Form; Figs. 6.8, 6.9). As already described, in complete clefts of the lip and palate, the lateral palatal segments are displaced laterally and the slopes of both

palatal segments are steeper than normal, with the palatal segments at the cleft space extending into the nasal chamber [5]. This steepness decreases with time, the slopes becoming more obtuse under the influence of tongue force. In clefts of the lip and palate, uniting the cleft orbicularis oris-buccinator superior constrictor muscle ring or using external facial elastics re-establishes the outer compressive muscular forces. This change in the muscle force vectors causes the laterally displaced palatal segments to move together. Moreover, this reduction in the width of the cleft is not limited to the alveolar process but extends as far back as the tuberosities of the maxilla and perpendicular pterygoid processes. The surgeon is challenged to establish muscle balance without disturbing the growth potential of the bony tissue being manipulated and to avoid scars that will tie or bind down the normally expansive forces of growth.

5.3 Reversing Aberrant Cleft Facial Forces in the Neonate

5.3.1 Lip Surgery, Elastic Traction, or Presurgical Orthodontic Treatment (Figs. 5.1–5.4)

1. Lip Surgery creates sufficient forces to bring the overexpanded palatal segments medially narrowing the alveolar and palatal cleft spaces. The surgeon often does this in two stages: First a lip adhesion at 3–5 months followed by a more definitive lip/nose surgery, which is more artistic. A cupid bow and normal nostrils are the eventual goals. (see Chap. 15)



Fig. 5.1 a–f. The use of an external elastic force to reduce premaxillary protrusion. **a** and **b** The protruding premaxilla extends forward in the facial profile. **c** Head bonnet with attached elastic placed against the protruding premaxilla causes it to ventroflex with the fulcrum at the premaxillary vomerine suture. **d** and **e** Facial photographs at 3 years of age. The lateral lip elements are united with the medial positioned prolabium over the protruding premaxilla in one stage. Because the premaxilla is already ventroflexed at the time of surgery, there is

reduced muscle tension at the suture sites. **f** Intraoral photograph shows excellent anterior and buccal occlusion even with bilateral deciduous cuspids in crossbite. Comment: A severe overbite or overjet with a buccal crossbite at this age does not create a functional dental problem or inhibit palatal growth. Midfacial protrusion is expected and even desirable at this age. A straight profile in the mixed dentition usually indicates a concave profile will develop in adolescence after the pubertal growth spurt

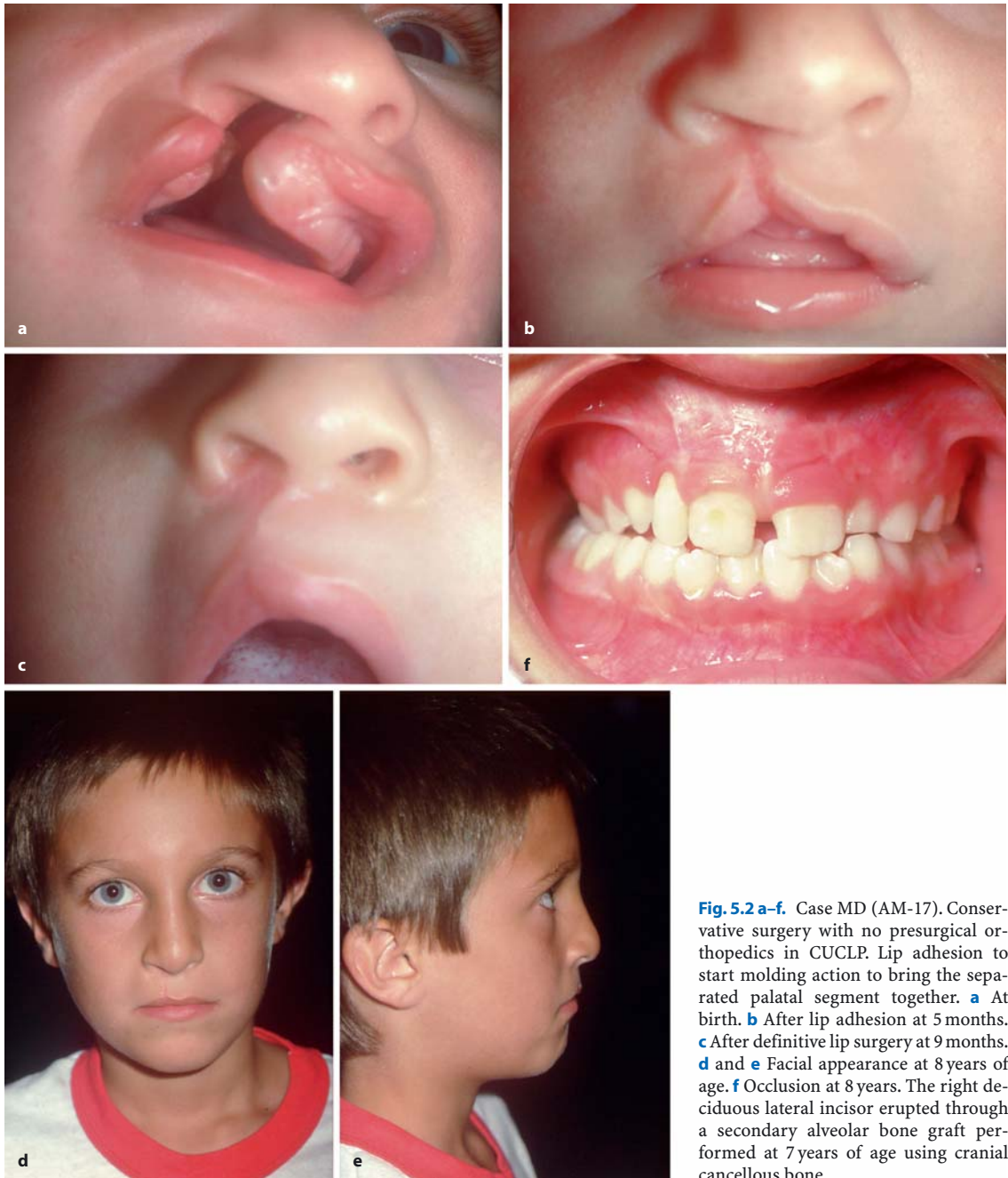


Fig. 5.2 a-f. Case MD (AM-17). Conservative surgery with no presurgical orthopedics in CUCLP. Lip adhesion to start molding action to bring the separated palatal segment together. **a** At birth. **b** After lip adhesion at 5 months. **c** After definitive lip surgery at 9 months. **d** and **e** Facial appearance at 8 years of age. **f** Occlusion at 8 years. The right deciduous lateral incisor erupted through a secondary alveolar bone graft performed at 7 years of age using cranial cancellous bone

2. Head Bonnet with Elastic Strap to be Placed over the Premaxilla in all Lip Clefts. The force system needs to be worn for 1 or 2 weeks along with arm restrains to prevent the infant overjet from removing the elastic strap. A premaxillary ventroflexion in CBCLP cases occurs very quickly creating an overjet and overbite. In CBCLP with a protruding

premaxilla at birth, the lateral palatal segments move medially behind the premaxilla. This relationship does not cause palatal growth retardation. Should a crossbite occur, the involve palatal segment usually can be moved laterally into proper occlusion at 4–6 years of age when the child is manageable in a dental chair.

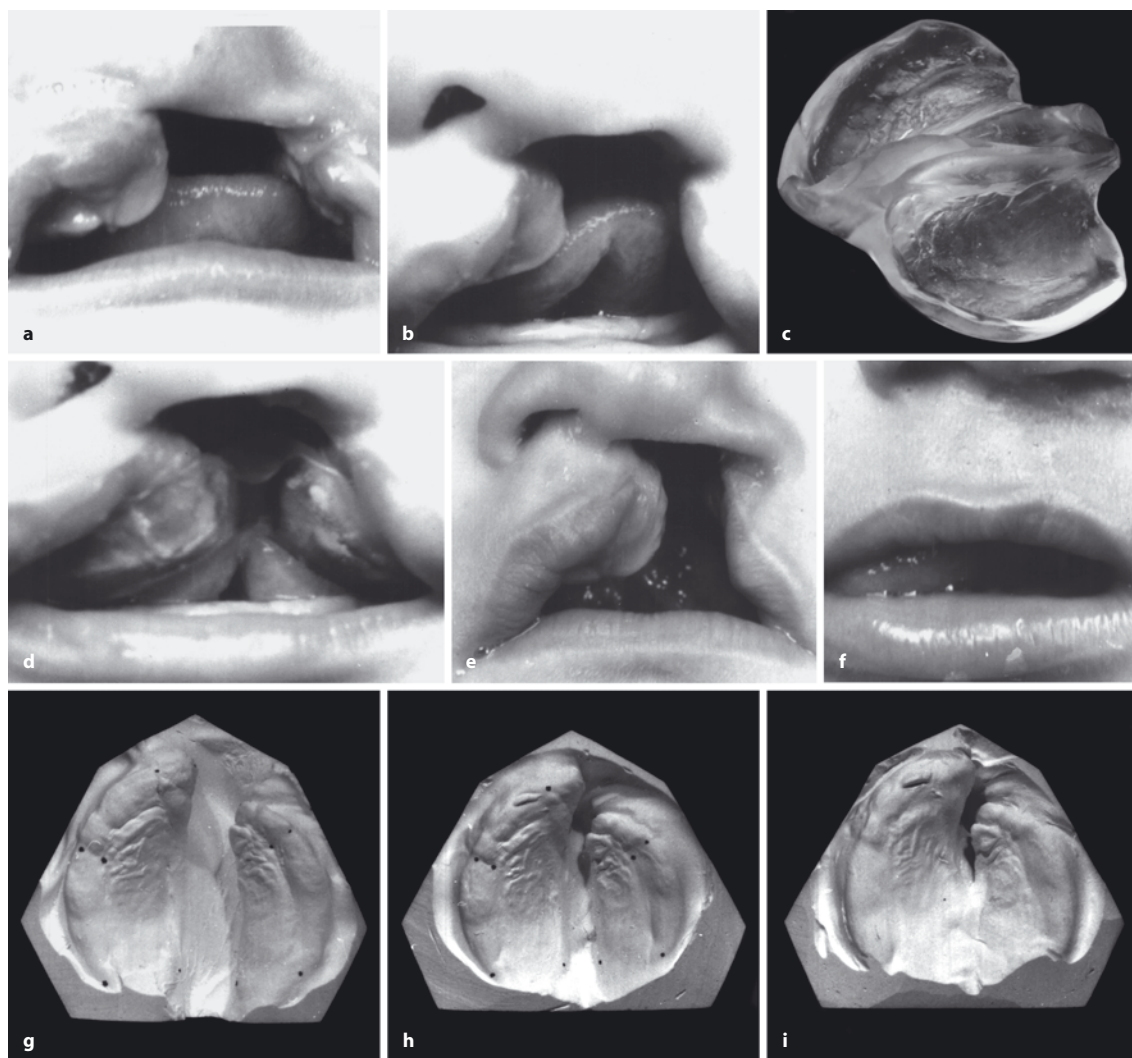


Fig. 5.3 a-i. Presurgical orthopedic treatment (PSOT) appliance for a CUCLP utilized from birth to 1 year 11 months at the University of Nijmegen (Courtesy of AM Kuijpers-Jagtman). **a** Lip and nose distortion at birth; **b** tongue posture within the cleft; **c** orthopedic appliance; **d** orthopedic plate prevents

the tongue from entering the cleft; **e** 15 weeks after PSOT and before lip closure; **f** 6 weeks after palate closure; **g** 17 months before soft palate closure; **h** at 14 months of age, before soft palate closure; **i** 8 weeks after lip closure

3. Presurgical Orthopedics: There are active and passive appliances, which are designed to create an alveolar butt joint [6]. In the distant past, primary bone grafting was utilized with the hope of stabilizing the palatal segment's position. However, with primary bone grafting it was found to cause midfacial deformity. Berkowitz in a recent longitudinal

palatal growth study [7] determined that the plates do not stimulate growth. Some surgeons who have used gingivoperiosteoplasty have created an anterior crossbite in most instances, which is hard to correct with expansion. Berkowitz strongly rejects the use of primary bone grafting and gingivoperiosteoplasty (see Chap. 20).

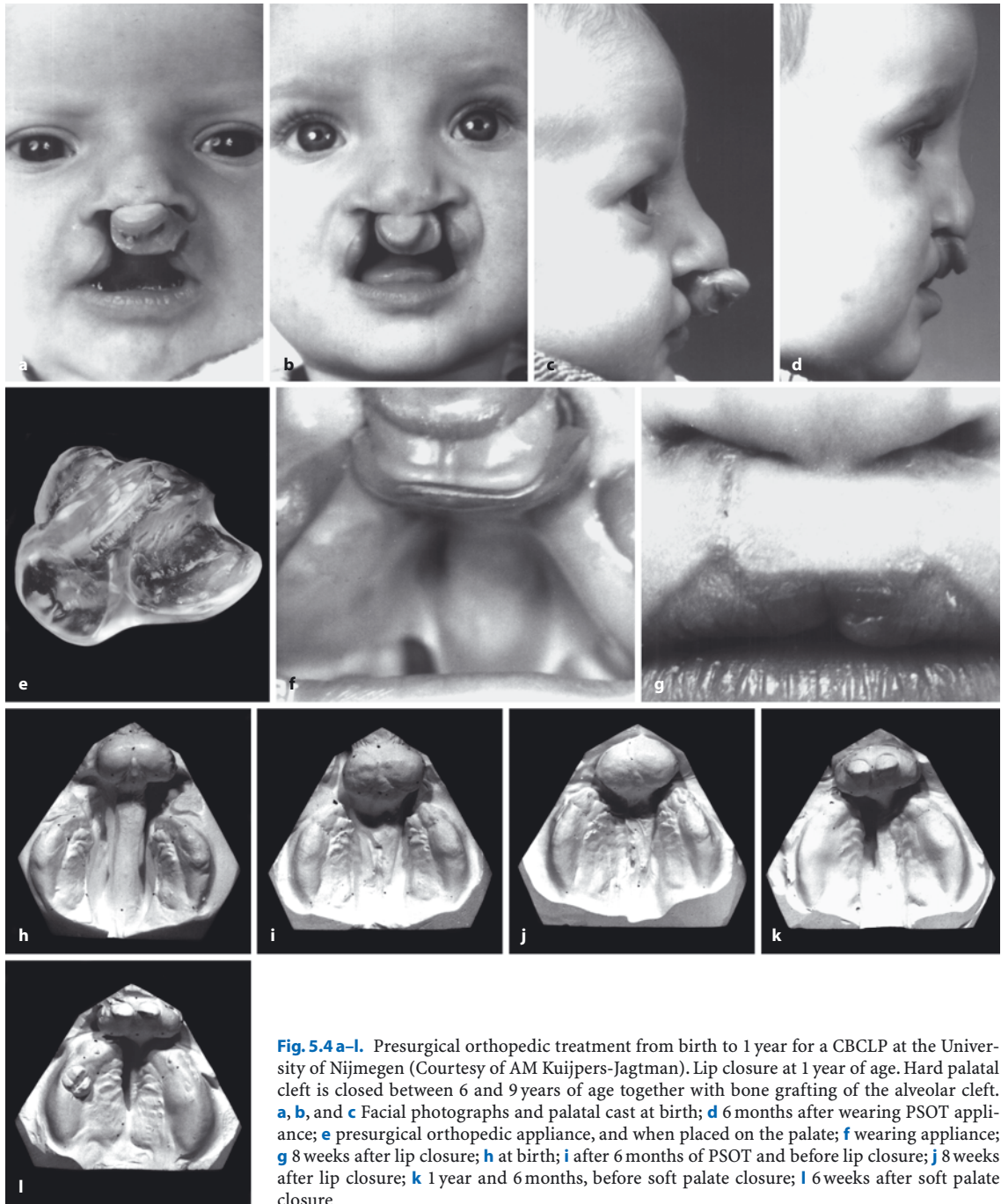


Fig. 5.4 a–l. Presurgical orthopedic treatment from birth to 1 year for a CBCLP at the University of Nijmegen (Courtesy of AM Kuijpers-Jagtman). Lip closure at 1 year of age. Hard palatal cleft is closed between 6 and 9 years of age together with bone grafting of the alveolar cleft. **a, b, and c** Facial photographs and palatal cast at birth; **d** 6 months after wearing PSOT appliance; **e** presurgical orthopedic appliance, and when placed on the palate; **f** wearing appliance; **g** 8 weeks after lip closure; **h** at birth; **i** after 6 months of PSOT and before lip closure; **j** 8 weeks after lip closure; **k** 1 year and 6 months, before soft palate closure; **l** 6 weeks after soft palate closure

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6

The Effect of Clefting of the Lip and Palate on the Palatal Arch Form

SAMUEL BERKOWITZ

6.1 Varieties of Cleft Lip and Cleft Palate

6.1.1 Action of Intact Facial Muscular Forces on the Maxillary Arch

In a normal jaw with intact lips and palate, the muscles of the lip, cheek, and pharynx exert their normal sphincter-like actions against the developing maxillary and mandibular arches. The compressive external muscular forces neutralize the expansion forces of the tongue (Fig. 6.1). The neonatal arch form changes as these forces change with growth and maturation; yet the opposing muscles always maintain a precise and dynamic balance with each other. When this muscular balance is upset, the arch form and teeth relationships change.

6.2 Aberrant Muscle Forces in Clefts of the Lip and Palate

A cleft of the lip and palate is the result of the failure of lip elements, and right and left palatal segments, to come together within the first 9 weeks of fetal life. The loss of muscular continuity of the orbicularis oris-buccinator-superior constrictor ring in complete unilateral and bilateral clefts changes the normal muscular force diagram. The aberrant muscular forces act to displace tissue masses. In complete clefts of the lip and palate, if the lateral palatal cleft segments are detached from the vomer, they will be pulled laterally by the external aberrant lip-cheek muscular forces, as well as spread apart by the tongue pushing into the cleft space (Fig. 6.2). Because clefts differ in their location and extent, lip and palate clefts can vary in the

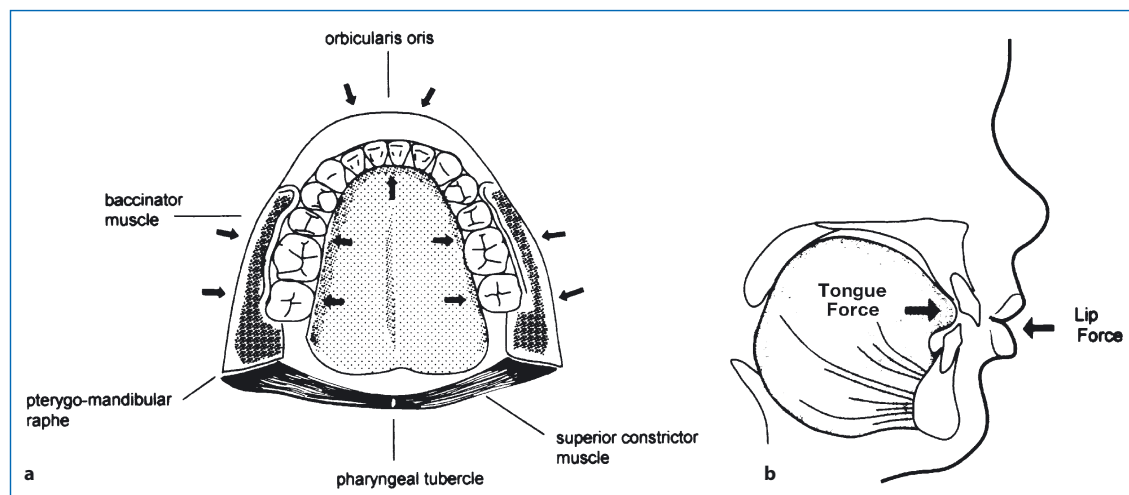


Fig. 6.1. The balance between facial and tongue muscle forces. **a** The outer muscles, the orbicularis oris, buccinator, and superior constrictor muscles, form a ring which acts to compress the palatal and mandibular arches with the teeth. **b** The tongue

force acts to expand the dental arches and teeth. Whether the teeth and arches align is determined by the resultant of these forces

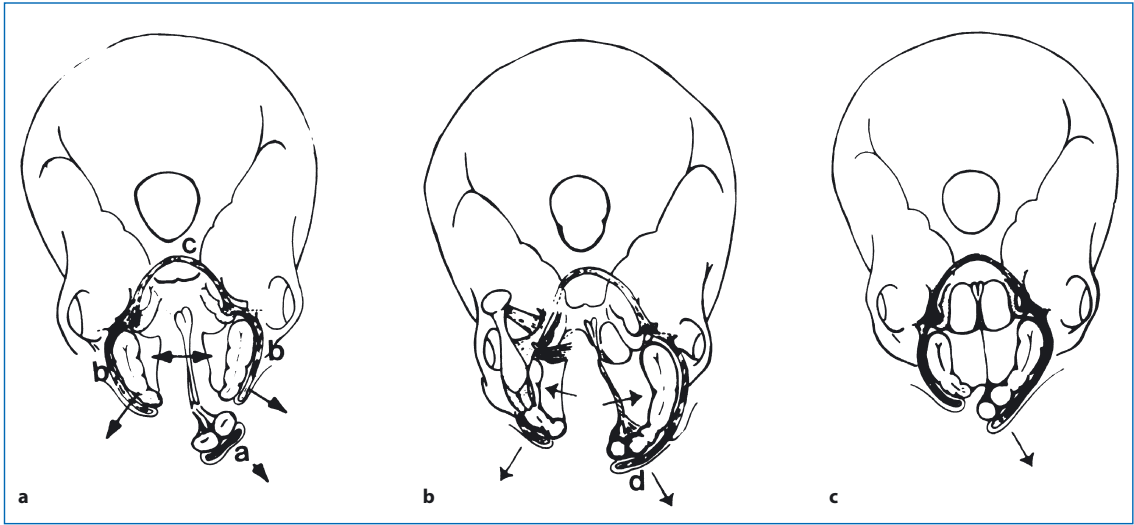


Fig. 6.2 a–c. Effects of complete clefts of the lip and palate at birth. In complete cleft lip and palate, with a separation in the orbicularis oris (**a**) -buccinator (**b**) -superior constrictor (**c**) muscle ring, the aberrant muscle forces plus the plunger action of the tongue causes the palatal segments to be pulled and pushed apart. **a** In bilateral clefts of the lip and palate, the premaxilla may be laterally or ventrally flexed with the fulcrum at

the premaxillary vomerine suture. **b** Complete unilateral clefts of the lip and palate at birth. The cleft's lesser segment and the premaxillary portion of the larger segment (**d**) are pulled outward. **c** Cleft of the lip and alveolus. The bony distortion is determined by the extent of alveolar involvement. (Courtesy of J. D. Subtelny.)

degree of geometric distortion, as well as in the size and shape of the cleft palatal segments.

The muscular forces that act on the bony scaffolding of the palate and pharynx begin very early in intrauterine life; therefore, the palatal and facial configuration at birth has been formed over the major portion of the infant's existence prior to birth.

In complete unilateral clefts of the lip and palate (CUCLP), the premaxillary portion of the noncleft segment is pulled antero-laterally. In addition to the lateral displacement of the lateral palatal segments, the premaxilla in the larger segment is carried forward in the facial skeleton. In complete bilateral cleft lip and palate (CBCLP), excessive growth in the

premaxillary-vomerine suture is caused by increased tension at this site, precipitated by mechanical force stresses during periods of rapid growth (Berkowitz [1], Pruzansky [2, 3], Friede [4, 5]) (Fig. 6.3). This growth is continuous during early postnatal years and provides a fourth dimension to the deformity, which can alter the cleft palatal segments and their associated parts and either simplify or complicate treatment.

If a soft tissue (mucous membrane, skin, and fibrous connective tissue), which collectively form Simonart's band, bridges the alveolar cleft, the attached palatal segments are limited in their degree of geometric displacement.

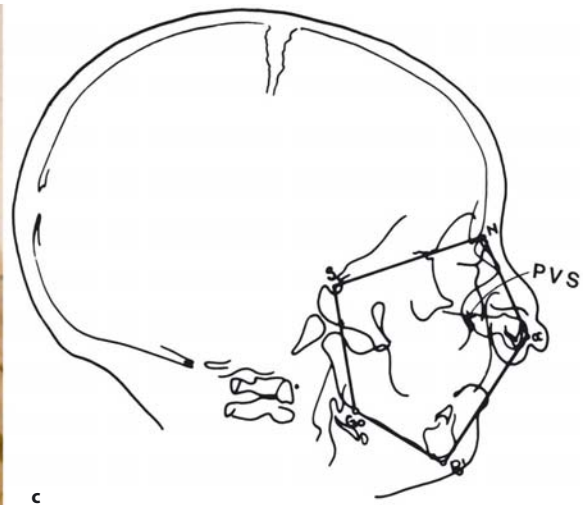
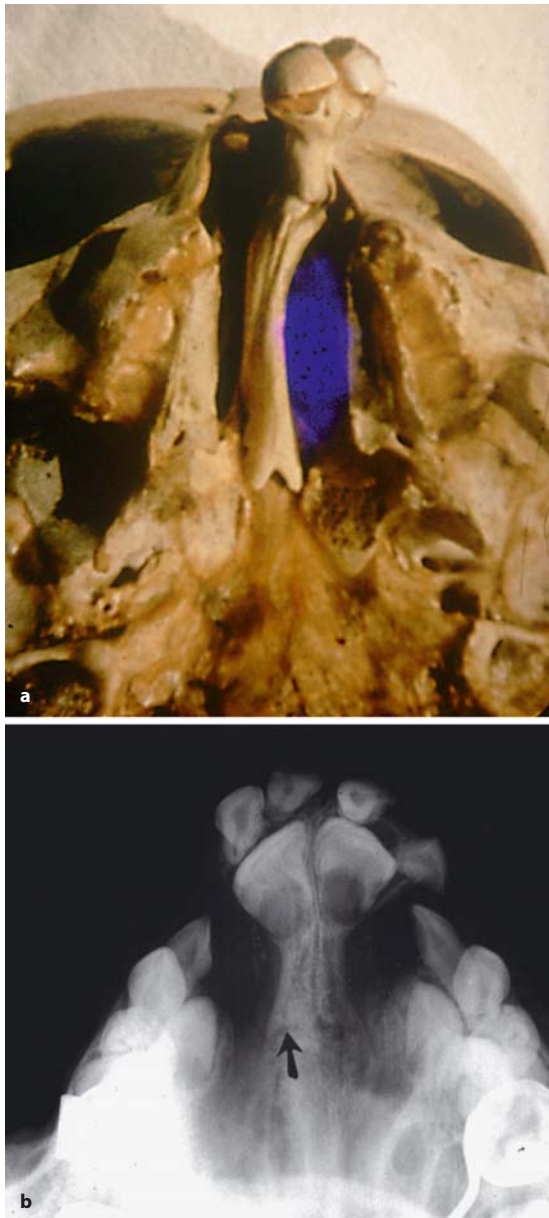


Fig. 6.3 a–c. Skull with a bilateral cleft and palate. **a** The palatal segments have been overexpanded and the premaxilla protruded by the resultant aberrant muscle forces. The premaxillary-vomerine suture separates the premaxilla and the vomer and is a growth site. **b** Lateral cephalometric tracing at birth. Lines connecting various landmarks create a polygon depicting their geometric relationships. This tracing of a CBCLP at birth shows the degree of premaxillary protrusion. *S* = Sella Turcica, *N* = Nasion, α = Alpha (the anterior extent of the premaxilla), *Po* = Pogonion, *Gn* = Gnathion, *Go* = Gonion, *PVS* = Premaxillary-vomerine suture. **c** Occlusal radiograph of the premaxilla and vomer showing the premaxillary vomerine suture (arrow)

6.3 Categories of Clefts

Depending on the elemental characteristics of the embryology, anatomy, and physiology of the cleft defect, the varieties of clefts of the lip and palate may be tabulated into four general categories: (1) those involving the lip and alveolus; (2) those involving the lip and palate; (3) those in which the palate alone is affected; and (4) congenital insufficiency of the palate. The term “palate” will include both the hard palate and the velum, or soft palate (Fig. 6.4).

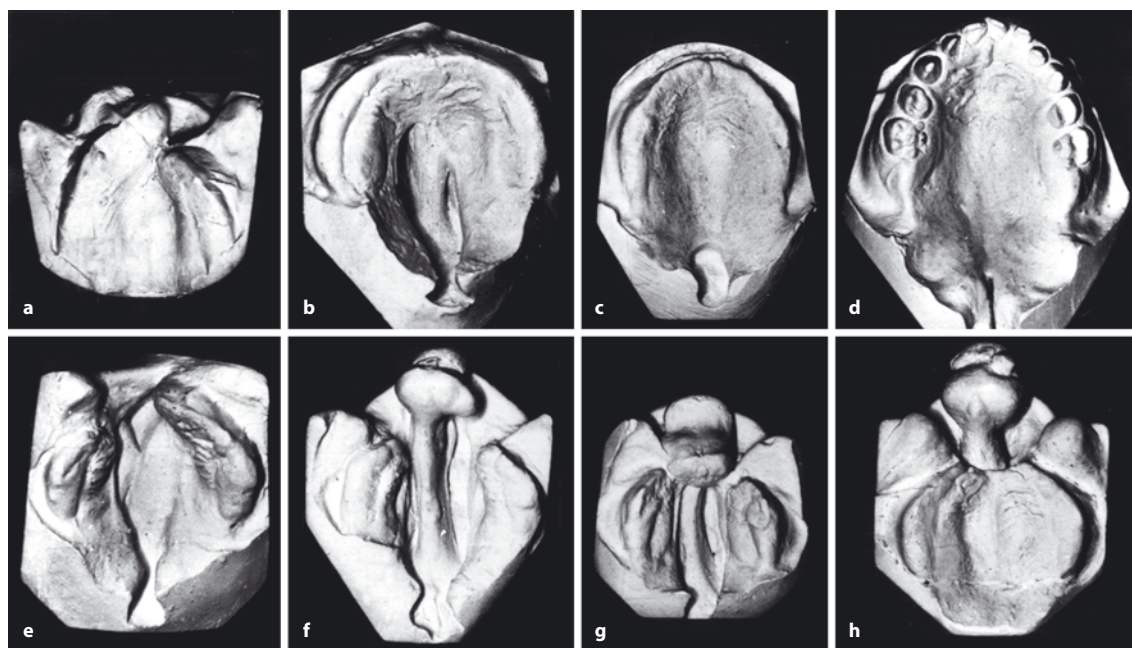


Fig. 6.4 a-h. The anatomic classification system is based on the location, completeness, and extent of the cleft deformity. Because the lip, alveolus, and hard palate develop from different embryonic sources, any combination of clefting can exist. **a** Cleft of the lip and alveolus. Normal palate. **b** Isolated cleft of the hard and soft palate. Normal lip and alveolus. **c** Cleft of the

soft palate and uvulae. **d** Cleft of the uvulae. **e** Complete unilateral cleft lip and palate. **f** Complete bilateral cleft of the lip and palate. **g** Incomplete bilateral cleft of the lip and palate. **h** Complete bilateral cleft of the lip and alveolus (Courtesy of Wolfe SA, Berkowitz S, eds. *Plastic Surgery of the Facial Skeleton*. Boston: Little, Brown and Co; 1989:292.)

6.3.1 Clefts of the Lip (Figs. 6.5 and 6.6)

A cleft of the lip may be complete, extending from the vermilion border to the floor of the nose, or it may be incomplete. There are various degrees of incomplete lip clefts. Minimal defects involving only the vermilion border are observed. In others, the defect may extend to the nose as a submucous cleft in the muscle band, bridged only by mucous membrane, skin, and fibrous connective tissue. The nasal alar cartilage on the side of the cleft is displaced and flattened to a greater or lesser degree, depending on the extent and width of the cleft. The tip of the nose is deviated toward the noncleft side.

The cleft in the lip may be unilateral or bilateral, occurring on one or both sides, respectively. If bilateral, it may be symmetrical or asymmetrical, that is, it may or may not involve the lip equally on both sides (Fig. 6.5b). It should be noted that, in bilateral clefts, a median portion of the lip is isolated in the midline and remains attached to the premaxilla and to the columella. This portion of the lip contains the philtrum. In complete bilateral clefts of the lip, the premaxilla protrudes considerably forward of the facial profile (Fig. 6.5b). It is attached to a stalk-like vomer and to

the nasal septum. The columella appears to be deficient, and the alar cartilages are flattened on both sides. The effect on the facial profile is to accentuate further the protrusiveness of the premaxilla and the portion of the lip which is attached to the facial surface.

The more complete the defect in the lip, the greater the influence of the cleft on the alveolar process. Because of this constant relationship between the lip and alveolar process, it is not necessary to include the alveolar process as a separate entity in this description and classification. The maxillary alveolar processes arise from the mesoderm in the depths of a sulcus separating the lip and palate, while the tegmen oris gives rise only to the soft palate and the central part of the hard palate.

The relationship between the degree of the cleft's effect on the alveolar process and defects in the deciduous and permanent dentition is interesting. The dental defect may be assessed in terms of the number of teeth, their shape, and structure as well as the position of the teeth in the dental arch. Irregularities in the alveolar process range from small dimples in association with minor clefts in the lip to actual grooves in the alveolar process to, in extreme cases of total clefts

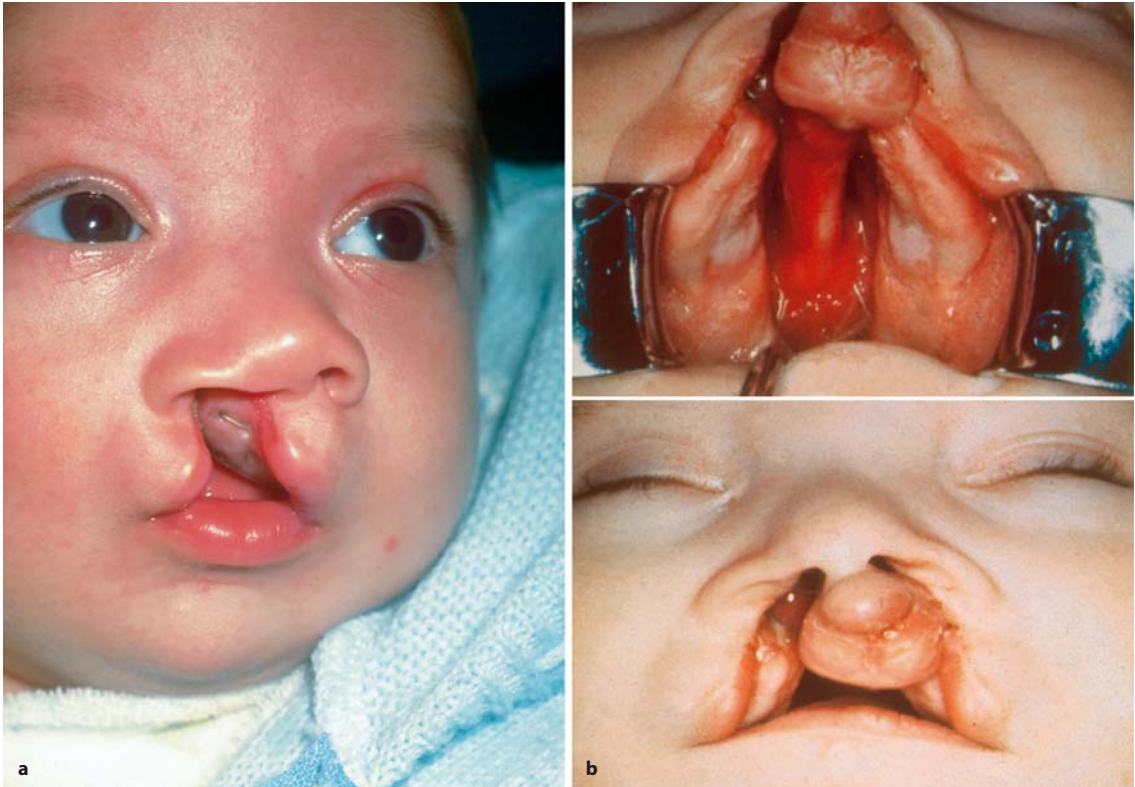


Fig. 6.5. **a** Complete unilateral cleft lip and palate. The distorted nostril is caused by the aberrant lip muscle forces. **b** Complete bilateral cleft lip and palate with a widely separated later-

al palatal segment. The protruding premaxilla extends forward of the lateral palatal segments and is attached to the vomer. The prolabium (central portion of the lip) overlies the premaxilla

Fig. 6.6 a,b. Incomplete clefts of the lip.
a Unilateral **b** Bilateral



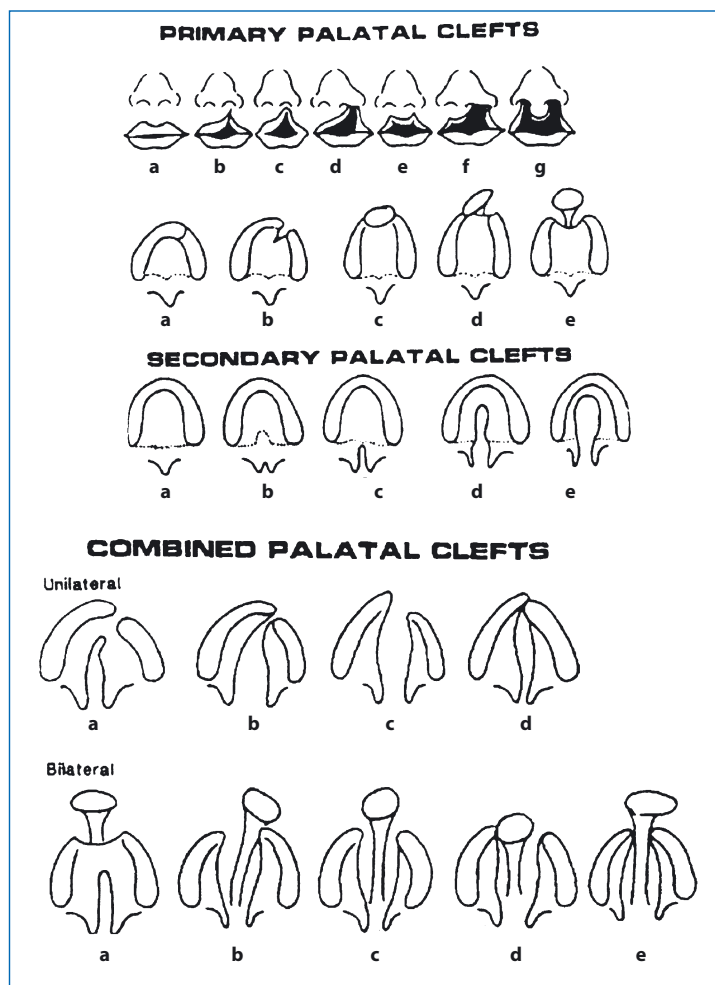


Fig. 6.7 a–e. Variations in the form, size, and extent of clefting in primary, secondary, and combined palatal clefts. Primary Palatal Clefts (with normal hard palate): Top row: **a** Normal lip; **b–g** The clefts may involve the lip only or may include the alveolus (tooth-bearing area) as well. The cleft can extend toward the nostril on one or both sides. Middle row, the cleft of the alveolus can extend to the incisal papilla on one or both sides to any degree. Bilateral alveolar clefts: **c**, incomplete on both sides; **d**, incomplete on one side and complete on the opposite side; **e**, complete on both sides. Secondary Palatal Clefts: **a** normal palate; **b** bifid uvula; **c** cleft of soft palate; **d** isolated cleft palate (moderate); **e** isolated cleft palate (extensive). Combined Palatal Clefts: Unilateral: **a** Isolated CP with cleft lip and alveolus; **b** Incomplete unilateral cleft lip and palate (IUCLP), cleft lip and alveolus are incomplete; **c** Complete unilateral cleft lip and palate (CUCLP); **d** Incomplete unilateral cleft lip and palate (IUCLP). Bilateral: **a** Complete bilateral cleft of lip and alveolus; **b** Bilateral-complete on one side, incomplete on the opposite with complete hard palate cleft; **c** Complete bilateral cleft of the lip and palate; **d** Bilateral incomplete alveolar cleft on one side, complete alveolar cleft on opposite side; **e** Complete bilateral alveolar clefts with both palatal segments attached to the vomer

in the alveolar ridge, displacement of the premaxillary segment toward the noncleft side. Small dimples or grooves in the alveolar ridge tend to fill in as the jaw grows. However, the deciduous lateral incisor that erupts in this area may be T-shaped, or otherwise misshapen, and malpositioned in the line of occlusion. Further documentation and analysis of serial records should provide detailed information concerning the eruption of teeth adjacent to the cleft in the alveolar process.

6.3.2 Cleft Lip and Cleft Palate (Fig. 6.7)

Clefts of both the lip and palate may be unilateral or bilateral. They may be complete or incomplete. In a complete unilateral cleft of the lip and palate, a direct communication exists between the oral and nasal cavities on the side of the palate where the cleft is situated. The nasal septum is attached to the palatal process on the opposite side, thus separating the nasal chamber from the oral cavity.

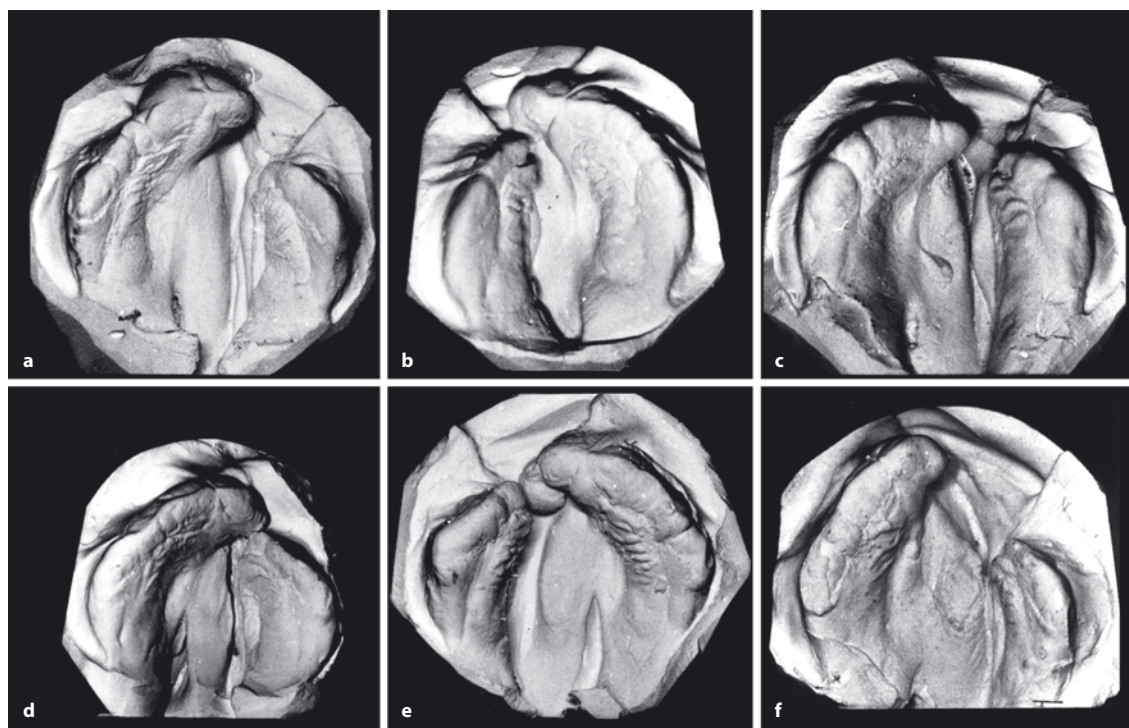


Fig. 6.8 a–f. Variations in unilateral clefts of the lip and palate at birth. The palatal segments may be complete (**a, c, e** and **f**) or incomplete (**b** and **d**); the cleft segment may be almost of the

same length or shorter. The cleft space may be relatively narrow (**b** and **d**) or wide (**a, c, e** and **f**)

A remarkable range of variation exists in each category. Indeed, various degrees of incompleteness of the cleft in the lip and palate may exist in combinations too numerous to describe conveniently. Moreover, some unilateral clefts of lip and palate exhibit wide separation of the palatal shelves. Others exhibit less separation, and in some cases, the segments actually overlap (Fig. 6.8). The palatal segment on the side of the cleft is often tilted medially and upward. The vomer is deviated from the midline at the line of attachment to the palatal process on the noncleft side. This deviation may be so extreme that the vomer assumes a nearly horizontal position at its inferior margin.

The bilateral cleft lip and palate also may be complete or incomplete (Fig. 6.9). If incomplete, it may be symmetrical or asymmetrical, depending on the equality of involvement on both sides. In the complete bilateral cleft lip and palate, both nasal chambers are in direct communication with the oral cavity. The palatal processes are divided into two equal parts, and

the turbinates are clearly visible within both nasal cavities. The nasal septum forms a midline structure that is firmly attached to the base of the skull, but is fairly mobile in front where it supports the premaxilla and the columella. Cephalometric roentgenograms reveal the existence of a suture line, the premaxillary vomerine suture, between the vomer and the premaxilla. This suture plays an important role in facial growth, and is also a point of flexion for the premaxilla upon the vomer (Fig. 6.3b and c).

The premaxilla may be small or large, symmetrical or asymmetrical. The number of incisor teeth contained in this segment is directly related to its size and shape. Permanent teeth may be missing and it may contain only 1 or more deciduous teeth when the cleft of the lip is complete on both sides, the premaxilla projects considerably forward from the facial aspect of the maxillae. This anterior protrusion is less evident if the lip is incompletely cleft on one or both sides.



Fig. 6.9 a-i. Variations in bilateral cleft lip and palate. The size of the premaxilla varies with the number of teeth it contains. Classification is dependent on the completeness of clefing of the lip and alveolus and whether there is a cleft of the hard and soft palate. Yet one or both sides of the hard palate may or may not be attached to the vomer. If it is attached to the vomer, it is classified as being incomplete. Even in complete clefts of the lip and alveolus the extent of premaxillary protrusion will vary. **a** Incomplete bilateral cleft lip and palate. Complete cleft lip and palate – left side. Incomplete cleft lip and palate – right side. **b** Complete bilateral cleft lip and palate. Complete cleft palate –

both sides. **c** Incomplete bilateral cleft lip and palate. Incomplete palatal clefts – both sides. **d** Complete bilateral cleft lip and palate. Incomplete right and complete left palate. **e** Incomplete bilateral cleft lip and palate. Incomplete left palate and complete right palate. **f** Complete bilateral cleft of the lip and palate. incomplete right and left palatal segments. **g** Complete bilateral cleft of lip and palate. Incomplete left palate and complete right palatal segment. **h** Complete bilateral cleft lip and palate. incomplete left palate and complete right palate. **i** Incomplete bilateral cleft lip and alveolus. Normal palate

6.3.3 Isolated Cleft Palate

In this defect, neither the lip nor the alveolar process is involved (Fig. 6.10). The cleft may involve only the soft palate or both the soft and hard palates, but never the hard palate alone. This observation is in accordance with the finding that fusion of the hard and soft palates proceeds from front to back (Fig. 6.11).

The cleft may extend forward from the uvula to varying degrees. In some cases, the cleft is limited to the uvula or to the uvula and soft palate. In others, it may extend into the hard palate. It is recommended that a digital examination of the posterior edge of the hard palate be performed. A midline notching will reveal the presence of a submucous cleft. The full extent of submucous clefts can be mapped by cephalometric

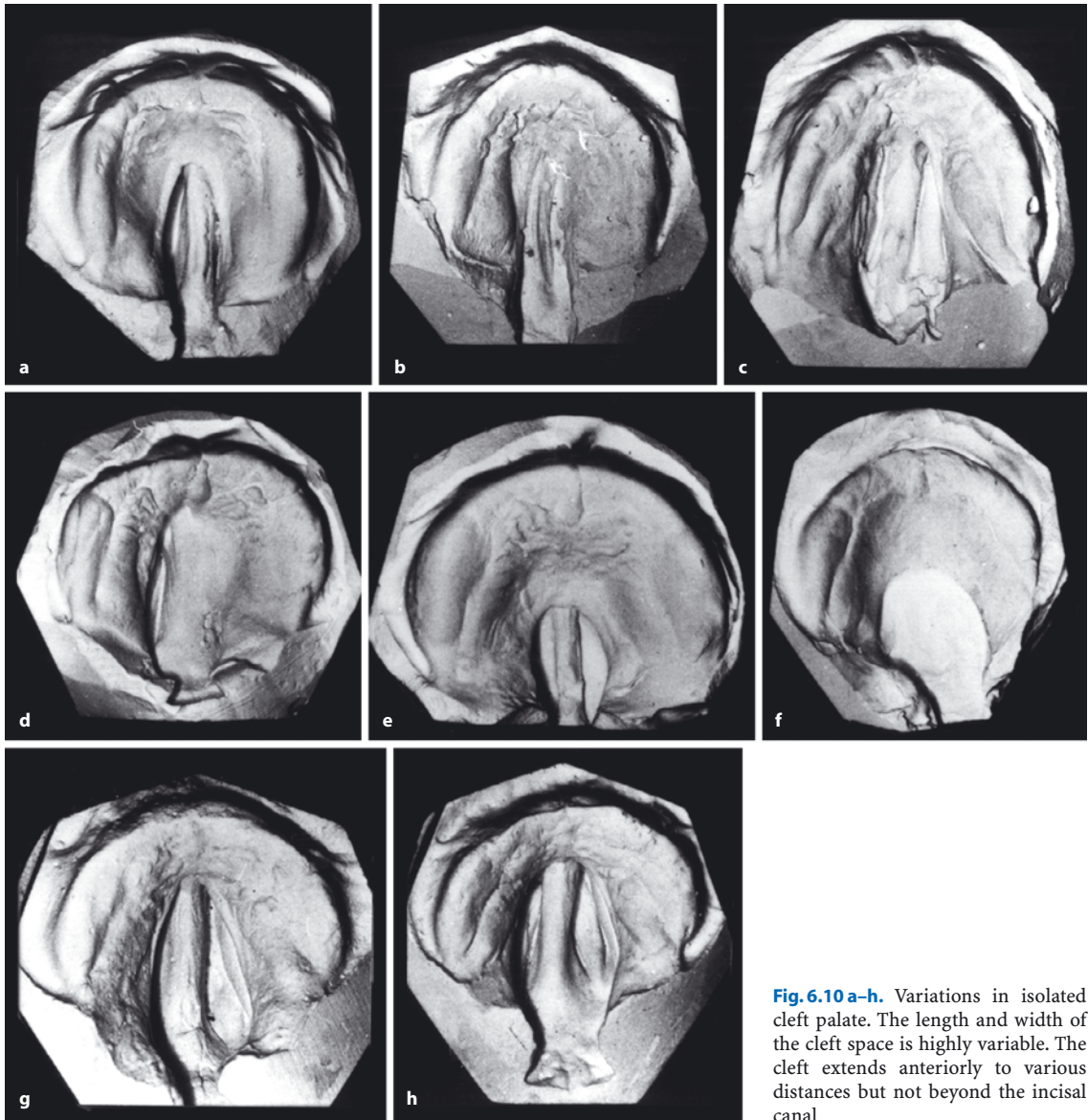


Fig. 6.10 a-h. Variations in isolated cleft palate. The length and width of the cleft space is highly variable. The cleft extends anteriorly to various distances but not beyond the incisal canal

laminagraphy or by transillumination through the nose.

In the extreme form, the cleft palate may extend anteriorly as far as the nasopalatine foramen, the incisal canal. When the cleft involves a considerable portion of the hard palate, the nasal chambers are in direct communication with the oral cavity. In most instances, the nasal septum is not attached to either palatal process throughout the extent of the cleft. However, occasional asymmetries may be noted in which the septum is attached to a portion of the palatal process on one side to a greater extent than to the palatal process on the opposite side.

The outline of the cleft may be wide or narrow, pyriform or V-shaped. Excessively wide dental arches often are associated with wide clefts that extend to a considerable degree into the hard palate. In such instances, the mandibular dental arch may be in complete lingual relation to the maxillary arch so that the cusps of the teeth do not interdigitate in occlusion.

Lateral cephalometric headplates reveal that the dorsum of the tongue, at rest, is elevated and postured within the nasal cavity. During deglutition the thrusting action of the tongue operates to separate the palatal processes. These abnormalities in the posture

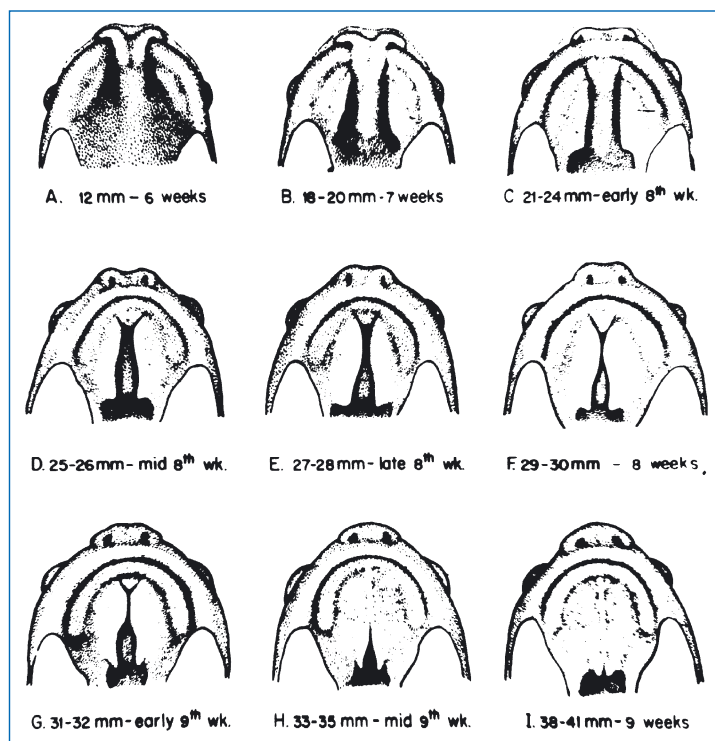


Fig. 6.11. Graphic summary of palatal fusion. (Redrawn from illustration in Kraus, Kitamura, and Latham. *Atlas of Developmental Anatomy of the Face*. New York, N.Y.: Hoeber Medical Division of Harper and Row; 1966.) The measurements and estimated ages given should be regarded as averages

and movements of the tongue are supported by the observations of speech pathologists. In this type of cleft, the vomer is significantly different in size and form from that observed in bilateral cleft lip and palate. In both types, the vomer is seen as a midline structure extending downward from the base of the skull. However, in bilateral cleft lip and palate, the inferior border of the vomer is thick and rounded, whereas in this category – cleft palate only – the vomer is thin and knife-edged. Serial observations reveal that the pattern of growth exhibited by the vomer is different in these two types of clefts.

Several other distinguishing characteristics apparent in some of the clefts in this category merit further comment. The high incidence of mandibular micrognathia found in patients with cleft palate gives credence to the theory that during embryonic development the tongue did not sink below the palatal processes, and thereby prevented their fusion in the midline. This raises the question of whether more than one causal mechanism might exist to produce the various kinds of clefts of the lip and palate.

In an extensive study of the mode of inheritance of cleft lip and cleft palate, Fogh-Andersen [6, 7] concluded that there are two different malformations with no genetic connection. In one group are clefts that involve the lip and occur most frequently in male patients. The other group is limited to clefts of the

palate, which are more frequent in female patients. According to Fogh-Andersen, the manner of inheritance differs for the two groups.

6.3.4 Submucous Cleft Palate (Fig. 6.12)

The classic triad of diagnostic signs is the bifid uvula, partial muscle separation in the midline with an intact mucosal surface, and the midline notch in the posterior edge of the bony palate. Hypernasality may or may not exist. Caution needs to be exerted prior to performing tonsillectomies and adenoidectomies because the velum may be functionally too short without the presence of the adenoid mass.

Berkowitz's cephalometric and nasopharyngoscopic studies have shown wide and unpredictable variability in the pharyngeal skeletal architecture and velar size and shape in submucous cleft palate as well as in all other cleft types (see Chapter 28). In some cases, due to a shallow pharyngeal space with relatively good velar length and mass with good lateral pharyngeal wall movement, no hypernasality existed. However, in most cases, the velum is usually too short, as well as too thin, and it fails to obturate the pharyngeal space properly. The problem appears to be due to an inadequate velum rather than a deficiency in lateral wall movements.

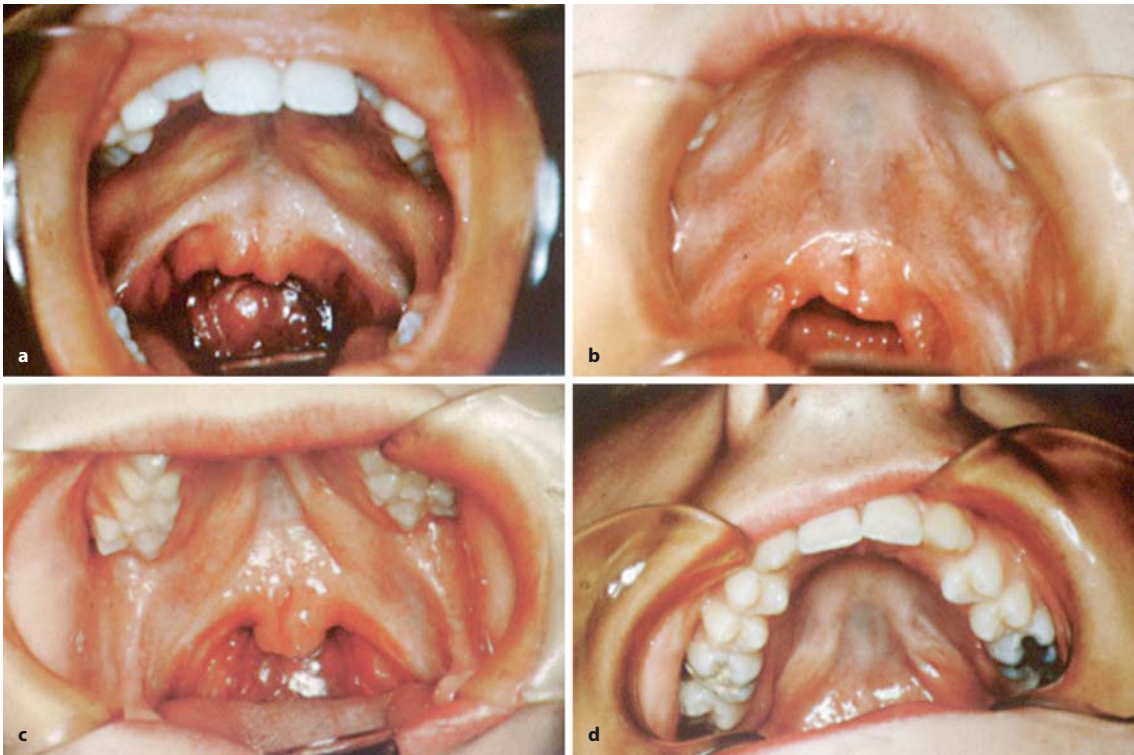


Fig. 6.12 a–d. Submucous cleft palate. This cleft is characterized by a bifid uvula, lack of muscle continuity across the soft palate, and a pink zone of mucosa (zona pellucida) across the

cleft in the hard palate. A palpable notch in the posterior border of the hard palate is always indicative of the presence of a cleft

The treatment of choice is a well-positioned and adequately wide superior-based pharyngeal flap. There is no apparent need to combine a palatoplasty with the pharyngeal flap. In patients with recurring infections of the adenoids, it is recommended that the adenoids be removed before the flap is placed. Speech therapy is an integral part of postoperative management.

6.4 Congenital Palatal Insufficiency (CPI)

It has been said that cleft palate is a type of defect that can be “seen, felt, and heard.” By contrast the defect known as congenital palatal insufficiency (CPI), until recently, has been more readily heard than seen or felt. This anomaly is seldom apparent at birth, and the first awareness of the defect occurs when the child develops the hypernasality characteristic of uncorrected cleft palate speech. The variety of factors producing this kind of speech defect can be examined by roentgenographic and nasopharyngoscopic methods (see Chapter 27).

Normally, during deglutition and during phonation, except for the sounds of “m,” “n,” and “ng,” the soft palate elevates, making contact with the posterior and lateral walls of the pharynx. The complicated synergies that contribute to this multi-dimensional contraction serve to separate the nasopharynx from the oropharynx. If for any reason this velopharyngeal closure cannot be achieved, deglutition is compromised, and in phonation the air stream necessary to create speech is misdirected through the nose. Palatal insufficiency may be caused by the velum being too short and/or by a deficiency in the antero-posterior dimension of the hard palate.

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6A Clefts of the Lip and Alveolus and Clefts of the Uvulae and Soft Palate

SAMUEL BERKOWITZ



6A.1 Clefts of the Lip and Alveolus (Figs. 6A.2–6A.4)

The alveolar portion is usually distorted outward in complete lip clefts. When the lip is united, the newly created lip force molds the alveolar section into proper alignment. A secondary alveolar bone graft is performed at the same age as in other cleft types (Fig. 6A.1).

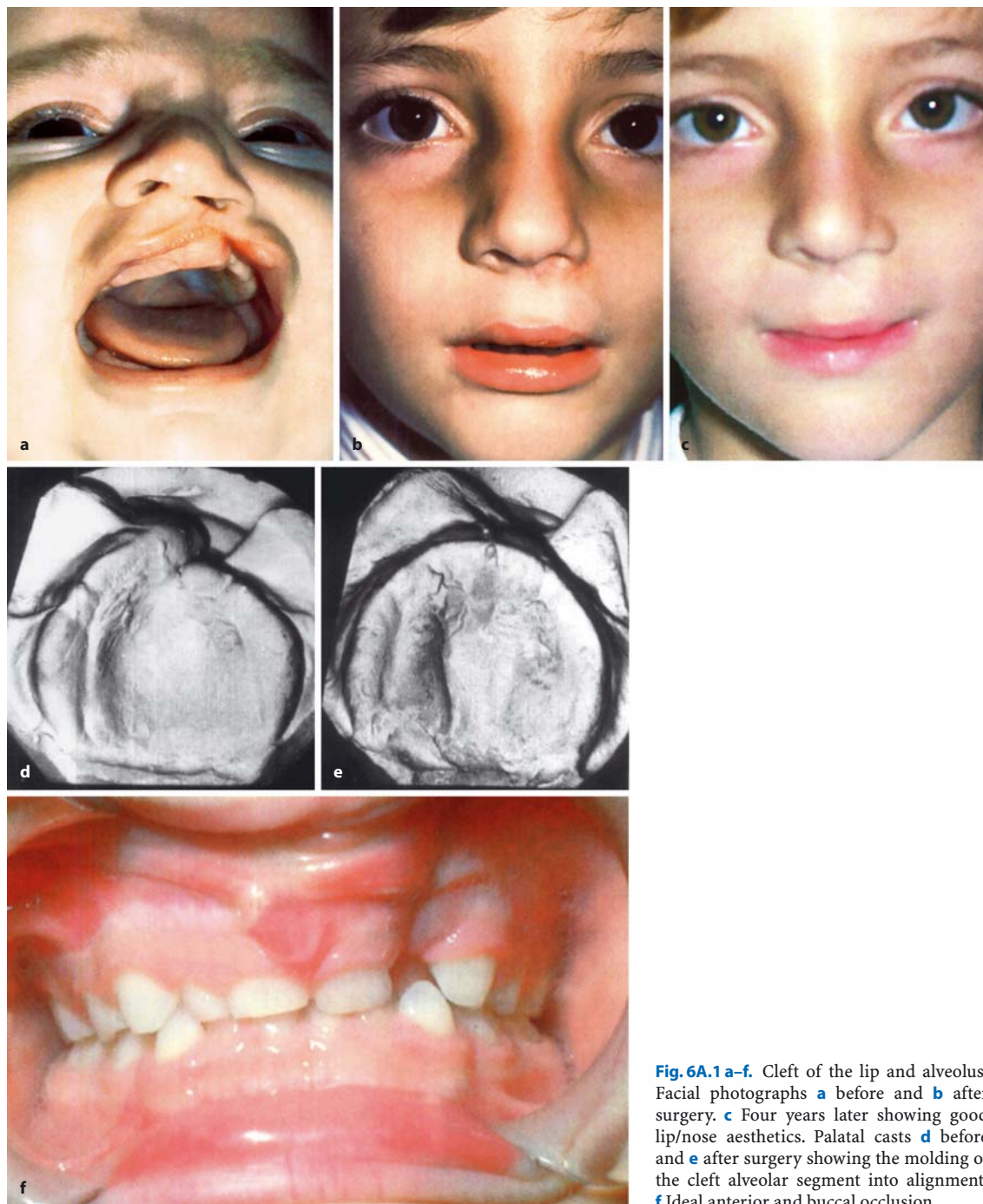


Fig. 6A.1 a–f. Cleft of the lip and alveolus. Facial photographs **a** before and **b** after surgery. **c** Four years later showing good lip/nose aesthetics. Palatal casts **d** before and **e** after surgery showing the molding of the cleft alveolar segment into alignment. **f** Ideal anterior and buccal occlusion



Fig. 6A.2. Case AS-30. Cleft of the lip and alveolus. Various lip revisions were performed



Fig. 6A.3. Case AS-30. Cleft of the lip and alveolus after orthodontics

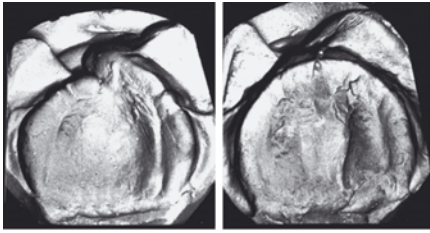
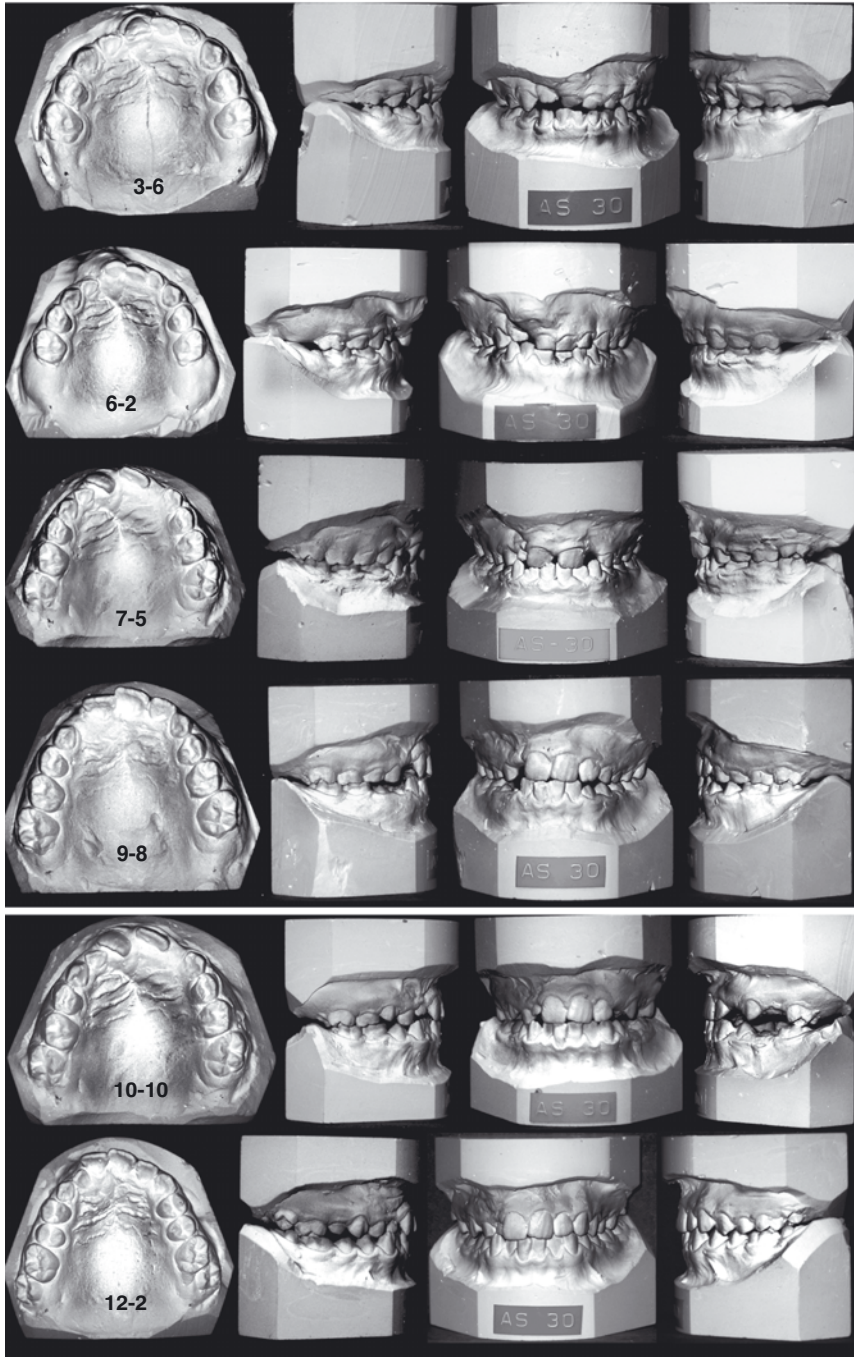


Fig. 6A.4. Case AS-30. Lip adhesion treatment and alveolar bone graft. Palatal growth and development was normal as seen by the occlusion



6A.2 Clefts of the Uvulae and Soft Palate (Fig 6A.5) and Cleft of the Uvulae Alone (Fig 6A.6)

When the health of the child permits, soft tissue clefts can be sutured within the first 3 months forces as Latham and his mentor McNeil have suggested. There are, however, some cases when, because of an unfavorable facial growth pattern coupled with a retruded maxilla relative to the anterior cranial bases, orthopedic protraction forces will be beneficial in the mixed (transitional) and permanent dentition.



Fig. 6A.5. Soft palate

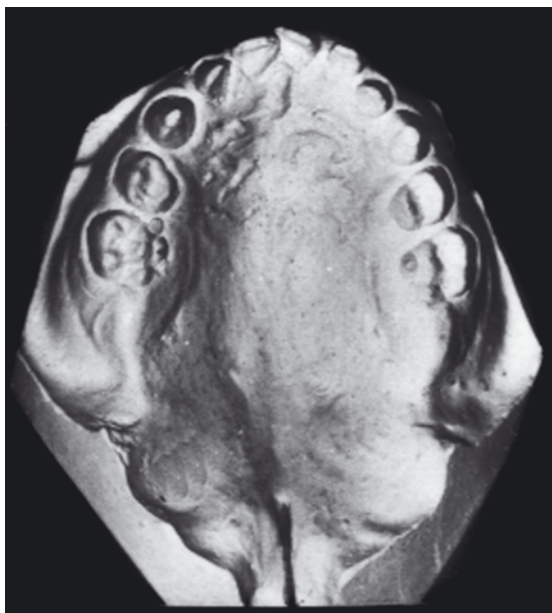
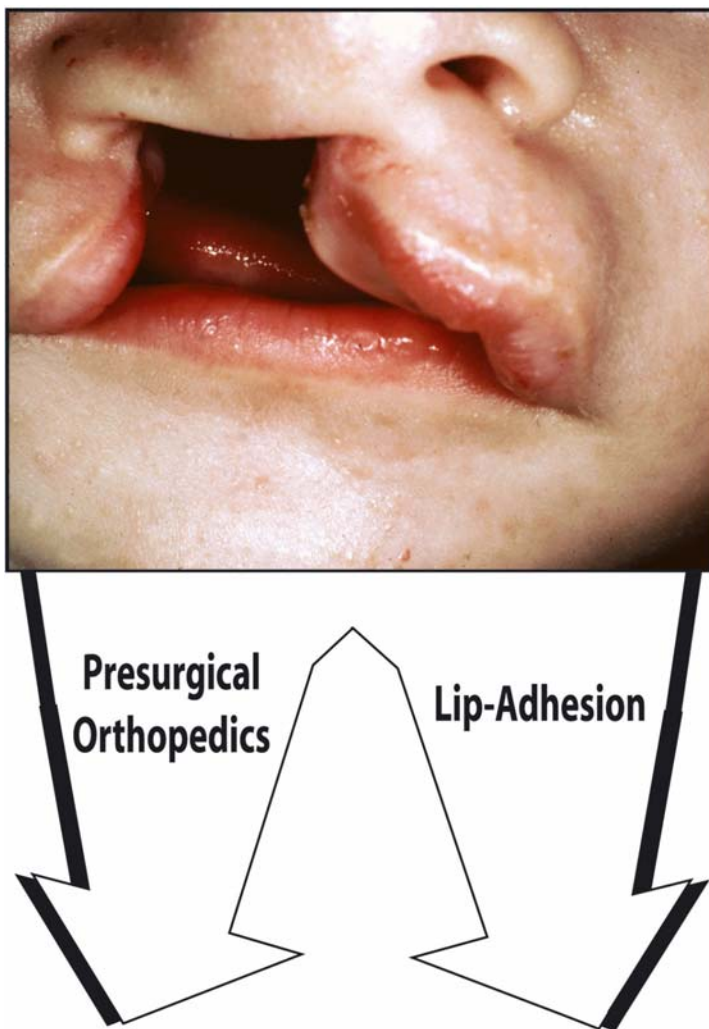


Fig. 6A.6. Uvulae

6B Complete Unilateral Cleft of the Lip and Palate

SAMUEL BERKOWITZ



As previously described [1], in complete unilateral and bilateral clefts of the lip and palate, after the lip is united, the overexpanded palatal segments move together, reducing the cleft width along its entire posterior length. Subtelný [2], using laminographs, has shown that newborns with complete clefts of the lip and palate have wider than normal pharyngeal widths, and the perpendicular plates of the sphenoids are distorted in their relationship. Aduss and Pruzansky [3] have demonstrated that, in complete unilateral cleft lip and palate, any one of three arch forms can result after the lip is repaired (Fig. 6B.1):

1. The alveolar segments can move into end-to-end contact, producing a symmetrical arch form.
2. The alveolar segments can overlap, producing what is erroneously known as a “collapsed” arch form.
3. The alveolar segments can move closer together but not make contact. This occurs because of an inhibiting factor of the inferior turbinate on the cleft side making contact with the distorted bulge of the nasal septum.

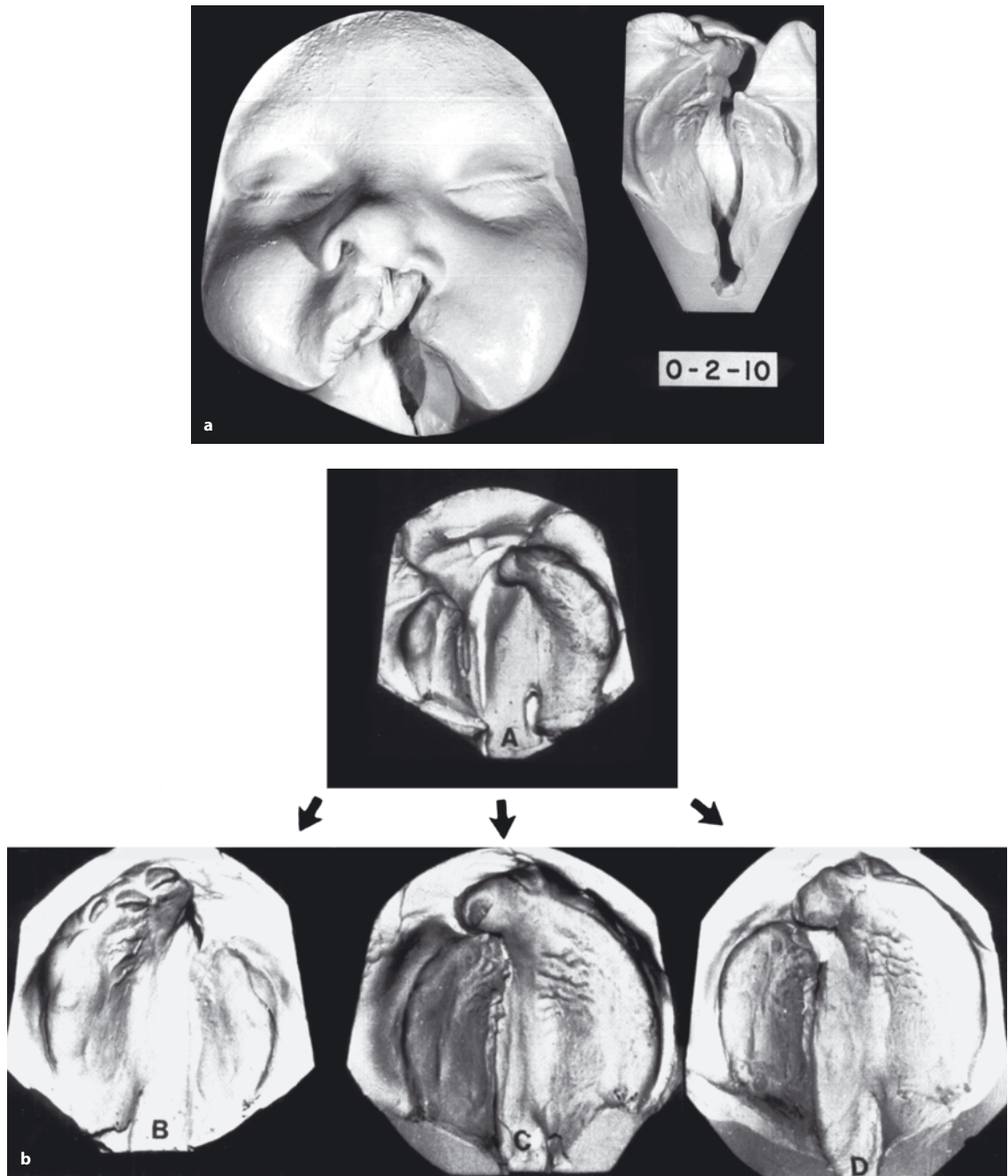


Fig. 6B.1. **a** CUCLP. Facial and palatal casts. **b** Complete unilateral cleft lip and palate (CUCLP) before (A) and after (B) lip surgery. With the establishment of muscle continuity, the lesser segment moves medially while the premaxillary portion of the larger segment moves medio-inferiorly, both acting to reduce the cleft width. Any of the following segmental relationships can result: **B** No contact between segments. The inferior turbinate on the cleft side makes premature contact with the

bowled nasal septum. **C** The premaxillary portion of the larger segment overlaps the smaller segment. **D** The segments form a butt joint showing good approximation. Pruzansky and Aduss have shown that there is no correlation between the original cleft width and the resultant arch form. Wider clefts seemed to demonstrate less of a tendency toward collapse than did the narrower clefts [3]

In a series of 58 patients who had no presurgical orthopedics or primary bone grafting, Aduss and Pruzansky [3] found that approximately 43% had overlap of the alveolar processes (mistakenly called collapsed arch). Among these patients, crossbites of the canine and first deciduous molar were the most common finding at 5 years of age. There were no anterior crossbites. Other investigators have reported similar results [4, 5]. Berkowitz [6], in a serial study of 36 cases with complete unilateral clefts of the lip and palate in which the lip had been united between the ages of 3 and 5 months and the palatal cleft closed between 18 and 24 months using a von Langenbeck with modified vomer flap without neonatal maxillary orthopedics, showed that 5 of the 36 cases had a complete buccal crossbite which was corrected within 6 to 10 months with fixed palatal expanders. Cuspid crossbite was the most frequent occurrence and was due to angular palatal rotation as well as to ectopic eruption of the deciduous cuspids. The cleft and noncleft segments were in either a Class I or Class II occlusal relationship. In no instance were any of the segments in a Class III relationship.

This confirms Berkowitz's belief that the cleft palatal segment is not repositioned within the skull relative to the mandible and that the maxillary-mandibular relationship is similar to that seen in the noncleft population. Whether the maxilla and mandible are both posteriorly positioned within skull has not been determined, others [7, 8] have found this to be the case. Therefore, the palatal segments do not need to be brought forward by the use of neonatal protraction forces as Latham [9] and his mentor McNeil [10] have suggested. There are, however, some cases when, because of an unfavorable facial growth pattern coupled with a retruded maxilla relative to the anterior cranial bases, orthopedic protraction forces will be beneficial in the mixed (transitional) and permanent dentition.

According to Aduss and Pruzansky [3], four factors govern arch form:

1. The size and shape of the alveolar process adjacent to the cleft. A bulbous and fully toothed alveolar process acts as an impediment to the collapse of the arch, whereas a thinly formed and dentally impoverished alveolar process leads to the overlapping of segments.
2. The size and shape of the inferior turbinate on the side of the cleft. A thick, rounded, well-modeled inferior turbinate can block excessive medial movement of the palatal segments.
3. The size and geometrical inclination of the nasal septum. A highly inclined septum with a contiguous bulbous turbinate will affect the movement of the palate and its final position.
4. The size and shape of the palatal shelves. Shelves of disproportionate size are more prone to overlap. One can certainly visualize that a long noncleft segment coupled with a short cleft segment will end up with the premaxillary portion overlapping the short cleft palate segment.

6B.1 Facial Characteristics

Aduss [11] in 1971 examined 50 males and 21 females with UCLP; their age range was between 4 and 14 years. He described craniofacial growth in the male cleft group as essentially equivalent to the female cleft group. He found that the gonial angle for the cleft patients was consistently larger than the noncleft group and the mandible appeared to be more retrognathic. He concluded that the craniofacial complex in the cleft sample tended to grow in a similar manner to that reported for the noncleft populations. The results of his study, based on a conservative method of surgery, negate the conclusions reached at the time regarding the deleterious effects of surgery on the growth of midface.

Hayashi et al. [12] studied craniofacial growth in unilateral complete clefts using lateral cephalograms of 135 males and 120 females with an age range of 4 to 18 years. Control subjects included 120 noncleft males and 120 noncleft females of similar age to the cleft subjects. They concluded that the cleft group differed from the control group in several major respects: (1) Their over-all growth trend showed a more downward or vertical direction; (2) the cranial base angle was more flattened; (3) the maxilla was smaller and was located in a more posterior and upward position; (4) ramal height was shorter, the gonial angle was more obtuse, and mandible was generally retrognathic; (5) upper face height was smaller and lower face height was greater; (6) underdevelopment in both the maxilla and the mandible was more pronounced in cleft females than in cleft males.

Smahel [13] in 1986, studied 30 boys with UCLP prior to palatoplasty using cephalometry. A comparison with 27 normal individuals matched in age showed that most basic deviations of the craniofacial configuration recorded in adults developed at an early age, often prior to palatoplasty, i.e., reduced height of the upper anterior face, maxillary dentoalveolar retroclination, displacement of the upper jaw backwards, widening of some components of the maxillary complex, and a shortening of the mandibular body and ramus. Only the length of the upper jaw was not reduced. The shortening of maxillary dimension occurred postoperatively at a more mature age.

Later in 1992, Smahel [14] presented another study of craniofacial morphology in UCLP in 58 adult males. The results showed a shortening of maxillary depth, reduction of the upper face height, increased lower anterior facial height and mandibular changes resulting from growth deficiency that consisted of shortening of the body and ramus, obtuse gonial angle, steep mandibular plane, and retrognathia.

Again in 1992 Smahel [14] studied growth and development of the face in UCLP during prepubertal and pubertal periods. He concluded that there were no definitive differences in the growth rate between the pre- and postpubertal periods. Therefore, the worsening of overjet during puberty could be due to the depletion of the compensation and adaptation after the previous orthodontic treatment rather than to the enhanced growth rate. In addition he found that, during the prepubertal period the lower jaw showed a very slight posterior rotation, while during puberty an anterior growth rotation was present. A marked retrusion of the maxilla developed already in the prepubertal period. During both periods there occurred an identical impairment of sagittal jaw relations and of the upper lip prominence, accompanied by a flattening of the facial profile and reduction of the nasolabial angle.

In 1996, Smahel [15] reported a longitudinal study regarding postpubertal growth and development of the face in UCLP as compared to the pubertal period. The data showed that in boys facial growth persists after the age of 15 years and maxillary growth attains almost half the values recorded in the period of puberty, while mandibular growth attains almost the same values as during puberty. In girls the growth is almost terminated except for the lower jaw, where it is still significant though several times slighter than during puberty. Due to the gender differences in the amount of postpubertal growth, developmental changes in facial configuration do not occur in girls during this period, while in boys there is a further deterioration of maxillary protrusion, sagittal jaw relations, and flattening of the face.

In 1988 Horswell [16] reported another long-term follow-up of skeletal growth of UCLP subjects ranging in age from 8 to 18 years. Serial cephalographs taken every 2 years were utilized for determination of six cephalometric dimensions: anterior cranial base, upper and lower facial heights, posterior nasomaxillary height, maxillary horizontal length, and mandibular length. These were compared to published cephalometric standards of a noncleft group. All dimensions except mandibular length were smaller in the UCLP group. The horizontal maxillary length appeared to be most affected in UCLP. Mandibular length was not affected in the cleft group.

6B1.1 The Oslo Study

Because of the stable and long history of meticulous record keeping and protocols that characterizes the data acquisition of the Oslo team, the following studies on unilateral cleft lip and palate are presented to provide a unique perspective on treatment strategies and facial growth standards based on longitudinal data. The author does not follow the same surgical strategies as those of the Oslo team, but recognizes that the differences are not significant enough to interfere with obtaining a successful long-term outcome.

Semb's [17] 20-year serial cephalometric study taken from the Oslo Archives gathered during Bergland's leadership involved 76 males and 81 females (157 individuals) who did not have neonatal maxillary orthopedics. All of the children in the study had lip closure in infancy using a modified Le Mesurier or, after 1969, a Millard procedure. During the same operation, the nasal floor was closed using a one-layer vomer flap. The remaining posterior palatal cleft was closed between 4 and 5 years of age using a von Langenbeck palatoplasty. Secondary alveolar bone grafts from the iliac crest were placed at between 8 and 11 years of age. By 1974 all palate repairs were completed by 18 months of age. Superior-based pharyngeal flap surgery for velopharyngeal insufficiency was performed in about 20% of the cases.

Compared with normal males and females, the pooled sample with unilateral cleft lip and palate showed: (1) skeletal and soft tissue maxillary retrusion; (2) elongation of the anterior face (even though the upper face height was shorter); (3) a retrusive mandible; (4) reduction in posterior face height; and (5) a slight increase in the angle of the cranial base.

The pattern of growth also was different from that of noncleft individuals. Between 5 and 18 years of age there was almost no increase in the length of the maxilla. There was a marked reduction in maxillary and mandibular prominence. Vertically, the excessive lower face angulations changed slightly.

6B1.2 Multicenter CUCLP Cephaloradiographic Study [18]

Ross's multicenter study involved data from 15 cleft palate centers around the world collected for the purpose of determining the effects of manipulative and surgical treatment on facial growth. A sample of 1,600 cephalometric radiographs of males with complete unilateral cleft lip and palate were traced, digitized, and analyzed in the Craniofacial Center of The Hospital for Sick Children. The seven series of studies con-

sidered virtually every aspect of treatment that might influence facial growth.

Ross concluded that the type of surgical repair used does not make an appreciable difference to facial growth. It appears, however, that there are differences that can only be explained on the assumption that some surgeons induce less growth inhibition than others. Variation of the timing of hard and soft palate repair within the first decade does not influence facial growth in the anteroposterior or vertical dimension. Ross admits that very early soft palate repair was not well represented in this study, and there is some suspicion that there might be untoward results.

Berkowitz's [19] clinical findings suggest a different conclusion that, in most cases, early surgery (before 12 months) will have a negative effect on palatal growth in all three dimensions. It all depends on the size of the cleft defect relative to the area of the surrounding mucoperiosteum. (see Chap. 17)

Ross's study did not include palatal surgery from 6 to 12 months. This study also reported that the resulting face is flat in profile and decreased in depth, with a vertical deficiency in the midface and vertical excess in the lower face. The mandibles in these faces characteristically are slightly shorter in total length so that the chin is retruded. The occlusion is more of a molar and incisor mesio-occlusion in clefts with less overbite and overjet. The soft palate in this sample is appreciably more posterior. The mandibular plane angle is greater, possible due to the need for more interincisal space.

Ross further stated that the bony pharynx was unaffected by treatment and that the variation in midface development can be attributed to maxillary length rather than to maxillary position. He also noted that the mandible is not directly affected by treatment. Facial growth is intrinsically compromised by an underlying deficit, and surgery acts to further interfere with growth of the midface by inhibiting forward translation.

The best results appear to follow lip repair at 4–5 months with no repair of the alveolus. Early alveolar repair restricts its vertical growth and should be avoided in individuals with poor growth potential. This leads to deficient midfacial height and poor vertical height proportions, with more acute nasolabial angles. There is no evidence that periosteoplasty will cause similar results. Berkowitz's [19] study conclusively shows that periosteoplasty inhibits midfacial development, especially that of the premaxilla (see Chap. 20)

Ross further states that the maxilla in the UCLP is not more posteriorly positioned to any appreciable extent, but it is much shorter in length. The repaired lip affects the basal maxilla more than the alveolar process. Vertical development of the posterior maxil-

la is more deficient than the anterior part. The mandible is shorter with a steeper mandibular plane angle.

Hard and soft palate surgical repair procedures provide the greatest potential for inhibiting the maxilla in length, forward translation, and posterior height.

Kwon's [20] retrospective longitudinal study of the skeleto-facial growth in unilateral cleft lip and palate documented and evaluated the proportional cranio-facial growth horizontally and vertically in 14 UCLP patients of the ages 5–18 years by using modified Coben's Basion Horizontal analysis. There were three populations included in this study: the Eastman cleft group (sample size 24) and the Miami cleft group (sample size 23) served as patient group and the Bolton templates (ages 5–18) served as controls. Samples were divided into 4 age periods according to the chronological ages, and then the growth pattern of each period were evaluated and compared. A total of 301 images of lateral cephalograms were examined and digitized. These characteristics of the skeletal facial growth of the UCLP are summarized as: (1) there is no difference of posterior cranial base over time; (2) maxilla is positioned posteriorly relative to Basion (BA) during the early ages and is getting retrusive due to the deficient growth with time; (3) Upper Anterior Facial Height (UAFH) is almost the same as the control; (4) Lower Posterior Facial Height (LPFH) is increased but is not as much as Lower Anterior Facial Height (LAFH); (5) Lower Anterior Facial Height (LAFH) is significantly increased; (6) Total Facial Height (TFH) is significantly increased; (7) mandible is positioned backward and downward due to the posterior position of the maxilla and the elongation of LPFH and LAFH; (8) skeletal profile is more convex and is getting straight and finally is flatter over time in the clefts than in the controls. Generally, the manifestation of the cleft characteristics of the Miami group is increased when compared to that of the Eastman cleft group. The skeletal growth leads to not only the maxillary retrusion but also to position the mandible down and back. Early orthopedic intervention followed by the fixed edgewise appliance and prolonged retention is recommended to try to correct the skeletal problems; camouflage by dental correction, and maintain to the treatment outcome with reasonable retainer.

6B1.3 Reflection on Ross' Excellent Multicenter Study [21]

In the Foreword of the multicenter study, Treatment Variables Affecting Facial Growth in Complete Unilateral Cleft Lip and Palate, Bruce Ross discussed the dif-

ficulty of performing this type of study due to the variability in sample size, age, sex, precise cleft type, and ethnic origin. He then mentioned the problems associated with doing cephalometric measurements and suggested using one center to control measurement errors; this was an excellent solution. According to Ross [21], the study considered virtually every aspect of treatment that might influence facial growth. An attempt was made to control many variables that influence growth research, so that a clear picture of the effects of each procedure would be available. Two major assumptions about the study are necessary if any conclusion can be drawn from these studies. The first is that all groups of infants with complete unilateral cleft lip and palate have exactly the same facial morphology at birth in spite of enormous individual variation within the group. The second assumption is that one group of infants will respond on the average in exactly the same way as any other group to a particular treatment. The intent was to assemble relatively pure samples of individuals who had received the given management techniques used consistently on all subjects from a particular center.

Berkowitz believes the study was a noble attempt by excellent clinicians/researchers to pool their sample cases to investigate treatment results. By necessity, it was limited to cephalometric records. By lumping all CUCLP cases together, regardless of the degree of palatal deformity at birth, much potential prognostic information for the treatment of individual cases is unavailable. Ross had no choice for discounting Slavkin's [22] and Ross and Johnston's [23] statements that palatal defects may be caused by either the failure of the separated palatal segments to fuse or, possibly, palatal osteogenic deficiency, a variable that needs to be considered in treatment planning. This statement on the embryo-pathogenesis of cleft palate explains why all clefts within a cleft type are not alike. It is not hard to reason that, as the extent of the cleft palatal defect varies, so will the resulting quantity of palatal surface area and the resulting quantity of postsurgical scar tissue. Because excessive scarring inhibits palatal growth and development, the palatal surface area at the time of closure needs to be considered in treatment planning. Berkowitz believes that the variability of palatal surface area within a particular cleft type weakens the value of Ross's [21] conclusions, which are based on the second assumption that "one group of infants will respond on the average in exactly the same way as any other group to a particular treatment." Berkowitz concludes that the next level of treatment evaluation studies designed to improve differential diagnosis requires the establishment of specific criteria based on quantitative and qualitative characteristics of the palatal defect when related to treatment outcome. (see Chap. 17)

6B.2 How the Palate Grows

As ready discussed in Chap. 4 on facial growth, bone growth involves the increase in size as well as remodeling. Serial palatal 3 dimensional growth studies by Berkowitz [19] has shown that growth and remodeling occurs over the entire palatal surface even at the medial border the palate at the cleft. (Figs. 6B.2, 6B.3)

6B.3 Treatment Sequence

6B3.1 Usual Treatment Sequence

1. Lip Adhesion: 3 months
2. Definitive Lip Surgery (Rotation Advancement): 10 months
3. Hard and Soft Palate Closure (von Langenbeck with vomer flap): 18–24 months (rarely 36 months).
4. Orthodontic Expansion (Quad Helix): 5–7 years.
5. Superior Based Pharyngeal Flap: 6–8 years if necessary.
6. Bone graft (iliac crest): 7–9 years.
7. Protraction Facial Mask (if necessary): 8 years or later.
8. Maxillary Surgical Advancement (Lefort I or Distraction Osteogenesis): varies.
9. Lip/Nose Revisions Techniques.

6B.4 Reports

In this section, treatment outcomes of selected cases are presented with photographs and dental casts. The casts started at birth and continued through adolescence; these records show the natural history of palatal and facial growth and development when conservative surgery was performed without the use of presurgical orthopedics.

In some cases, the lip was united after the use of a Logan's Bow (Fig. 6B.4), in others after lip adhesion at approximately 3 months of age. In some cases the cleft of the soft palate was united at the same time the hard palate was closed. Definitive lip surgery was performed at 6 months and hard palate closure using a modified von Langenbeck procedure with a vomer flap was performed between 12 and 24 months of age.

Selected cases are presented in Figs. 6B.5 through 6B.25 to show various treatment solutions to complex problems.

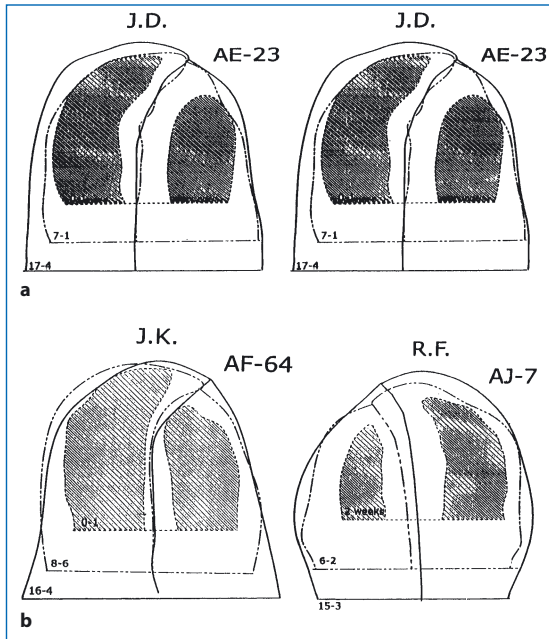


Fig. 6B.2 a, b. Superimposed computer generated images of serial CUCLP casts superimposed on the rugae and registered on the vomer AP line. The alveolar ridge is the outer limits of the palate. Surgery: Lip adhesion at approximately 3 months, definitive lip surgery at approximately 6 months and hard and soft palate closure between 18 to 24 months using a von Langenbeck procedure with a vomer flap. No presurgical orthopedics. Results: These 4 illustrations show the result of molding and growth. The least growth occurs anteriorly. Most of the growth occurs posteriorly to accommodate the developing deciduous and permanent molars. The palatal mucoperiosteum covers increase palatal size and the palatal cleft, which greatly reduced in size

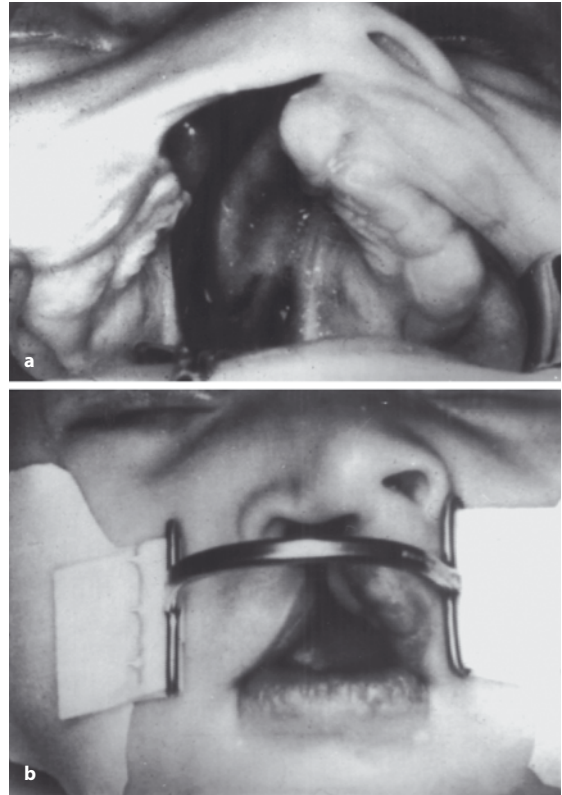


Fig. 6B.4 a, b. Logan's bow. Pressure is placed on the cheeks to bring the lips together prior to surgery. The bow helps to reduce tension at the suture line

Fig. 6B.3. Case KK-55. Serial growth of the palatal segments in CUCLP. Using computer-generated 3D images the surface areas mesial to the alveolar ridges were analyzed using an electromechanical digitizer. The same surgery stated in Fig 6B-2 were used. Results: This case is an example of 60 cases analyzed in the study; it shows: (1) Both palatal segments grow at the same rate, (2) the most rapid period (velocity) of growth occurs during the first 18 months. Comments: Because the most rapid period of growth occurs between 8 and 24 months when cells are most active, it is best to postpone palatal surgery until a later age in order to not inhibit growth

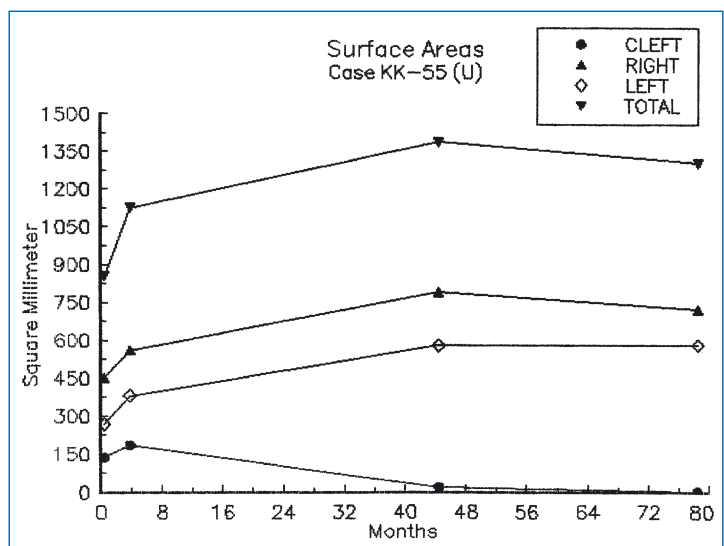




Fig. 6B.5 a–v. Case: KC (ZZ-1) demonstrates good palatal and facial growth in CUCLP. A very small cleft space at 5 months of age allowed for easy closure without much scar formation. Surgical treatment: No presurgical orthopedics. Lip adhesion followed by Millard's Rotation Advancement. Soft palatal closure at 2 months. Palatal left closure at 15 months using modified von Langenbeck procedure. Secondary alveolar cranial bone

graft at 6 years and 8 months. Photographs showing various treatment stages from birth to 17 years of age. **a** and **b** Newborn. **c** Lip adhesion at 4 months. **d** Lip at 2 years of age. Orthodontics during the deciduous dentition: **e** 2 years, showing anterior crossbite. **f** 2 years, 7 months: palatal view showing fixed buccal expander



Fig. 6B.5 a–v. (continued) **g** Anterior teeth were advanced and the cleft buccal segment expanded. **h** 5 years. Fixed palatal retainer. **i, j, and k** Fixed palatal retainer with lateral incisor pontic (tooth). **l** and **m** Facial photographs at 6 years

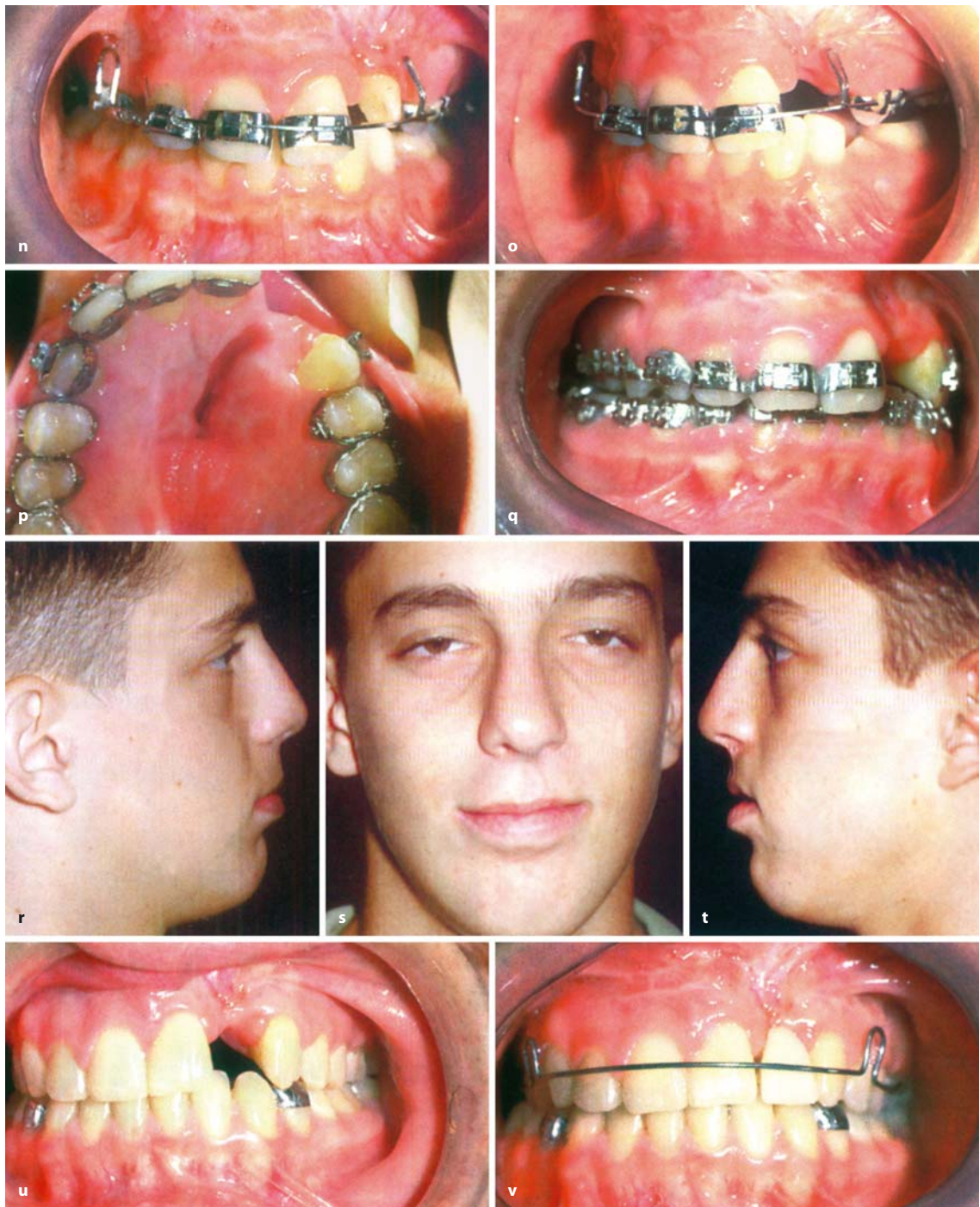


Fig. 6B.5 a-v. (continued) **n** and **o** 7 years, 3 months: lateral incisor is erupting through cranial bone graft. Orthodontics in the adult dentition: **o** Lateral incisor is extracted due to poor root development. **p** and **q** conventional orthodontics. Surgery

to close the palatal fistula was unsuccessful. **r**, **s**, and **t** Facial photographs at 17 years. **u**, and **v** Intraoral photographs. Hawley orthodontic retainer with lateral incisor pontic

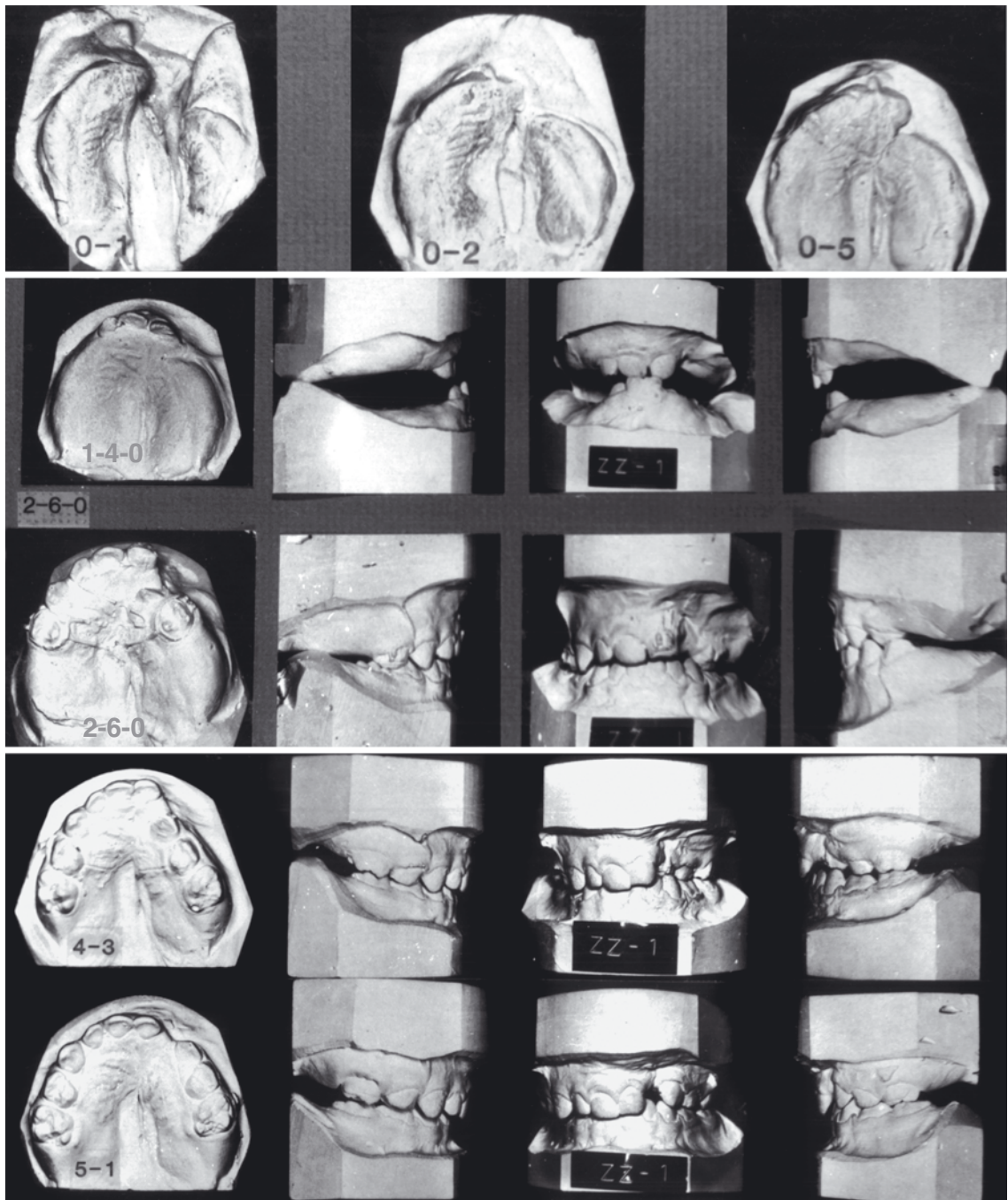


Fig. 6B.6. Case KC (ZZ-1). Serial casts from 0-1 to 0-5 shows medial movement and growth changes to the palatal segments. 0-5 The cleft space is extremely small with the palatal segments making contact anterior to the cleft space. 2-6-0 and 4-3

Mesioangular rotation of the lesser segment placed the deciduous cuspid in crossbite. 5-1 A fixed palatal expander rotated the segment outward, placing the teeth in ideal occlusion

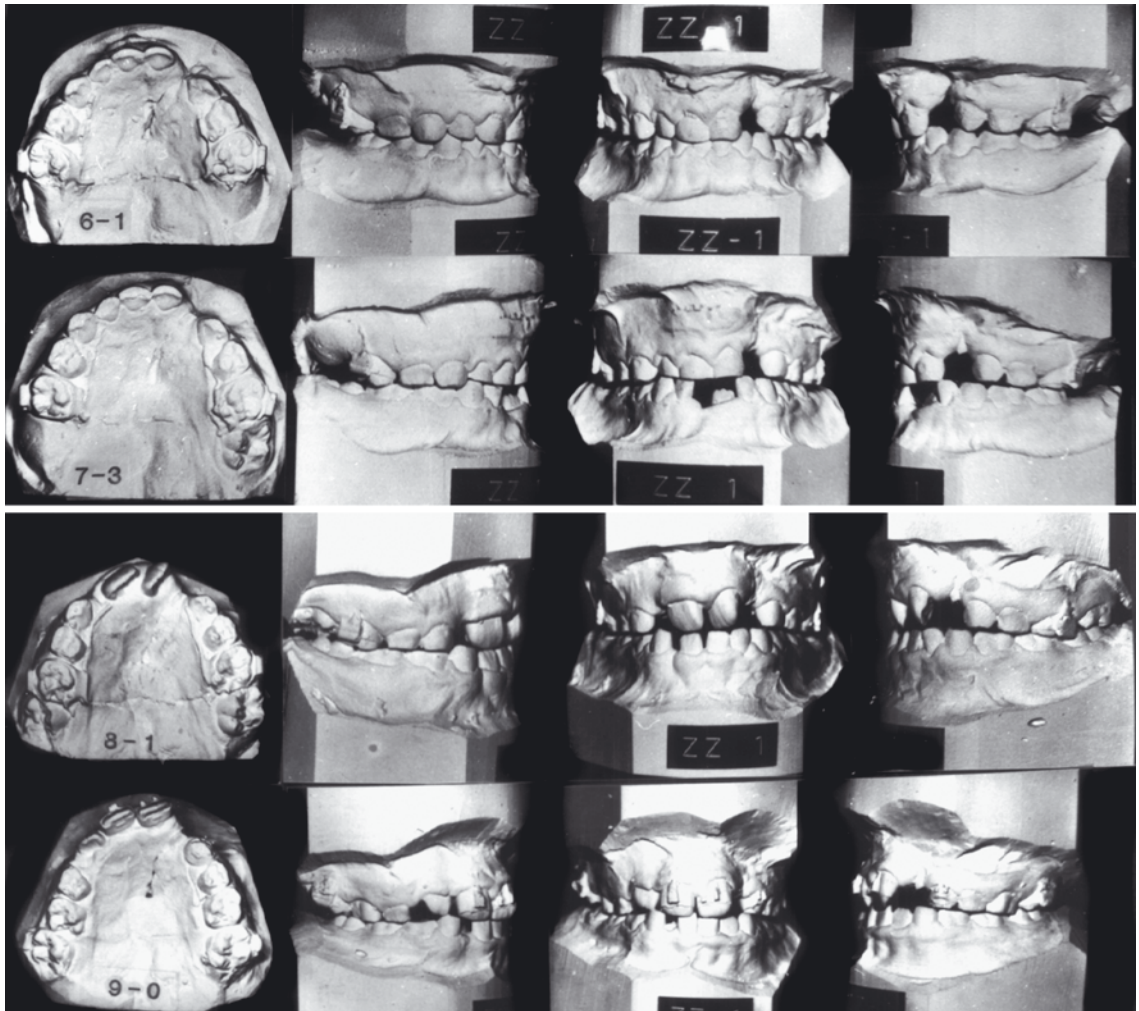


Fig. 6B.6. (continued) 6-1 Fixed retainer maintained the correction. Secondary alveolar bone graft was performed at 7 years, 3 months of age. 9-0 The maxillary anterior teeth were rotated for aesthetic reasons

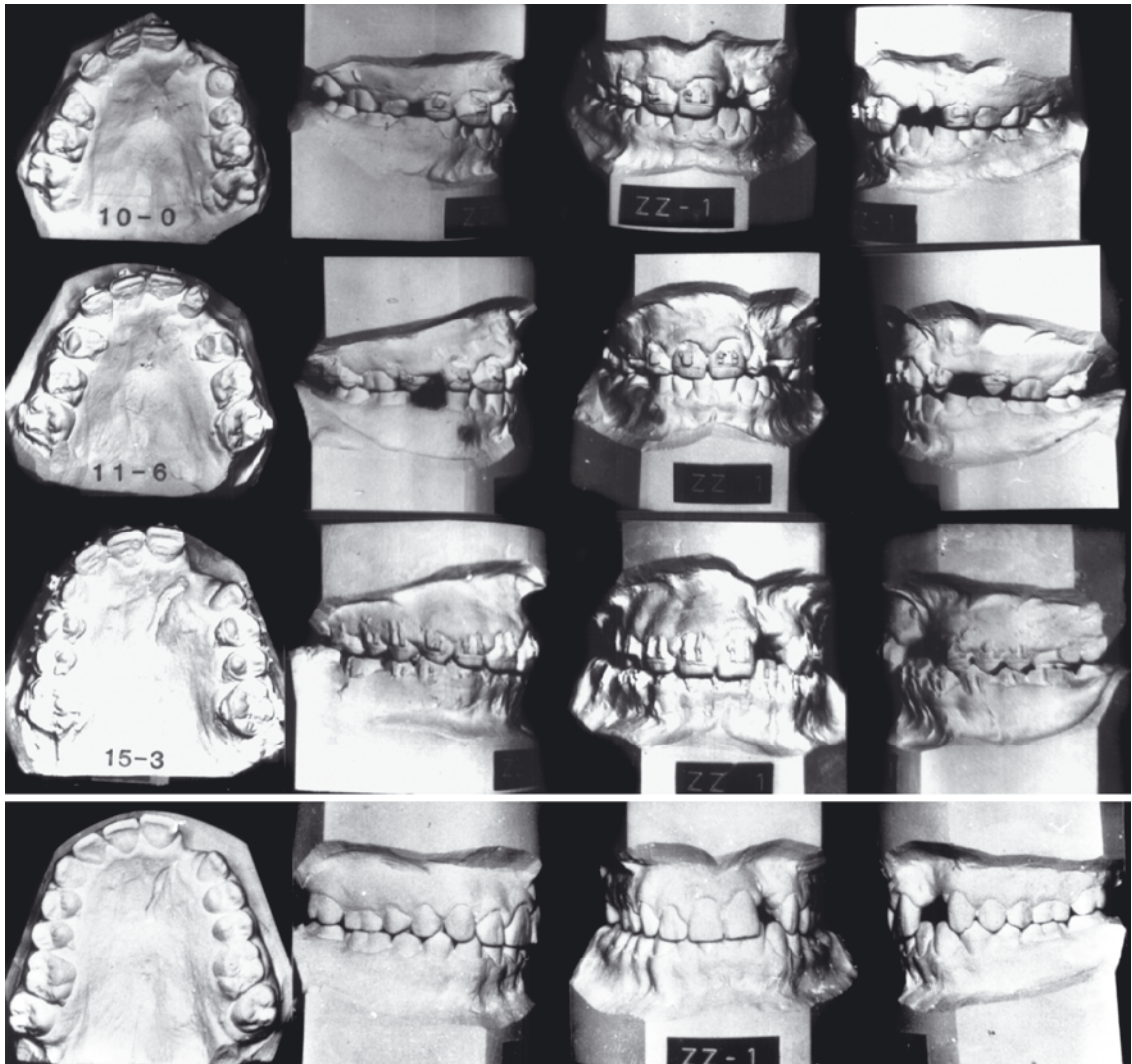


Fig. 6B.6. (continued) 11-6 The left lateral incisor is now in place within the arch. As a result of poor root development, it had to be extracted. Conventional orthodontics was instituted and completed by 15-3. Maxillary fistula was surgically closed at 16-3, and the arch form maintained with a removable Hawley retainer with a lateral incisor pontic. 17-0. Final occlusion. Comment: Because most cleft palatal arches have some degree

of osteogenic deficiency, when all bicuspid are retained, it is usual for the second molars to be blocked out and be impossible to position within the arch. This then necessitates their removal with possible replacement by the still unerupted third molars. In some instances, a small palatal fistula may not pose a speech problem or be a source of nasal drainage

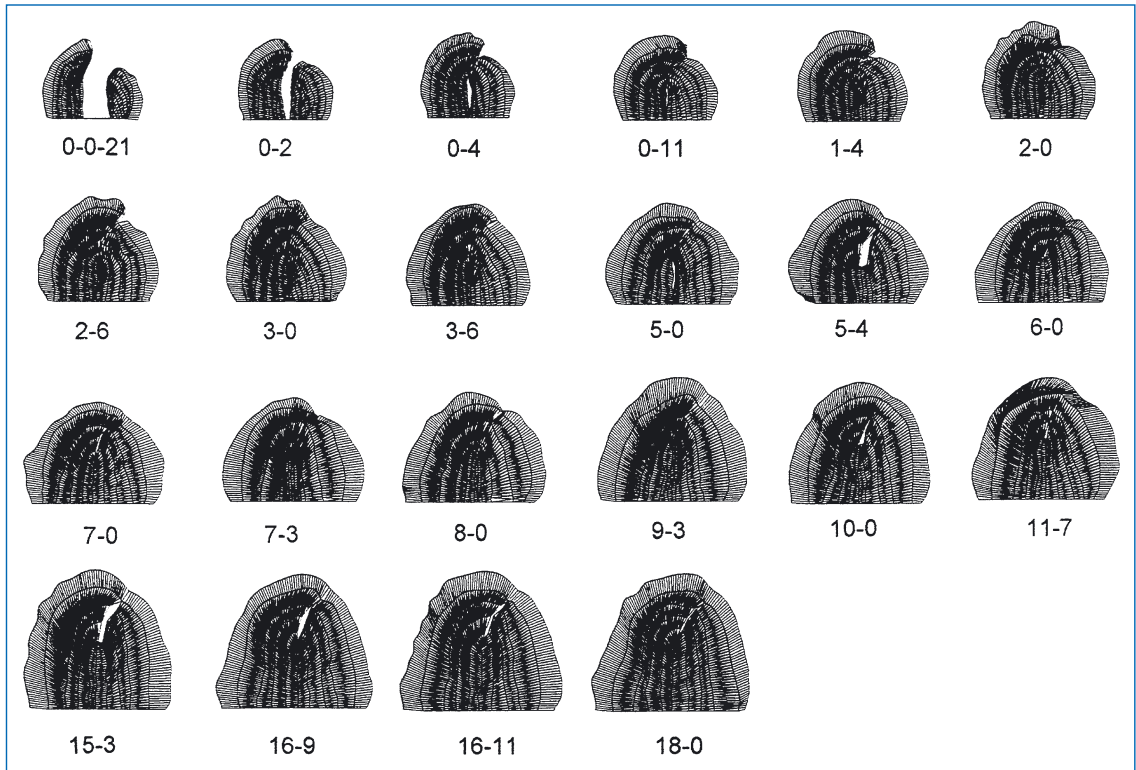


Fig. 6B.7. Case KC (ZZ-1). Computer-generated drawings of serial casts which are in the same scale. The soft palate was united at 2 months and the hard palate closed at 15 months. This series demonstrates a rapid reduction in palatal cleft size with molding action and palatal growth. A palatal “fistula” was ex-

posed when the cleft buccal segment was expanded to correct the crossbite. It was closed but reappeared when final orthopedic treatment moved the palatal segments slightly apart. The “fistula” did not penetrate into the nasal chamber. Therefore, it did not pose a speech or feeding problem

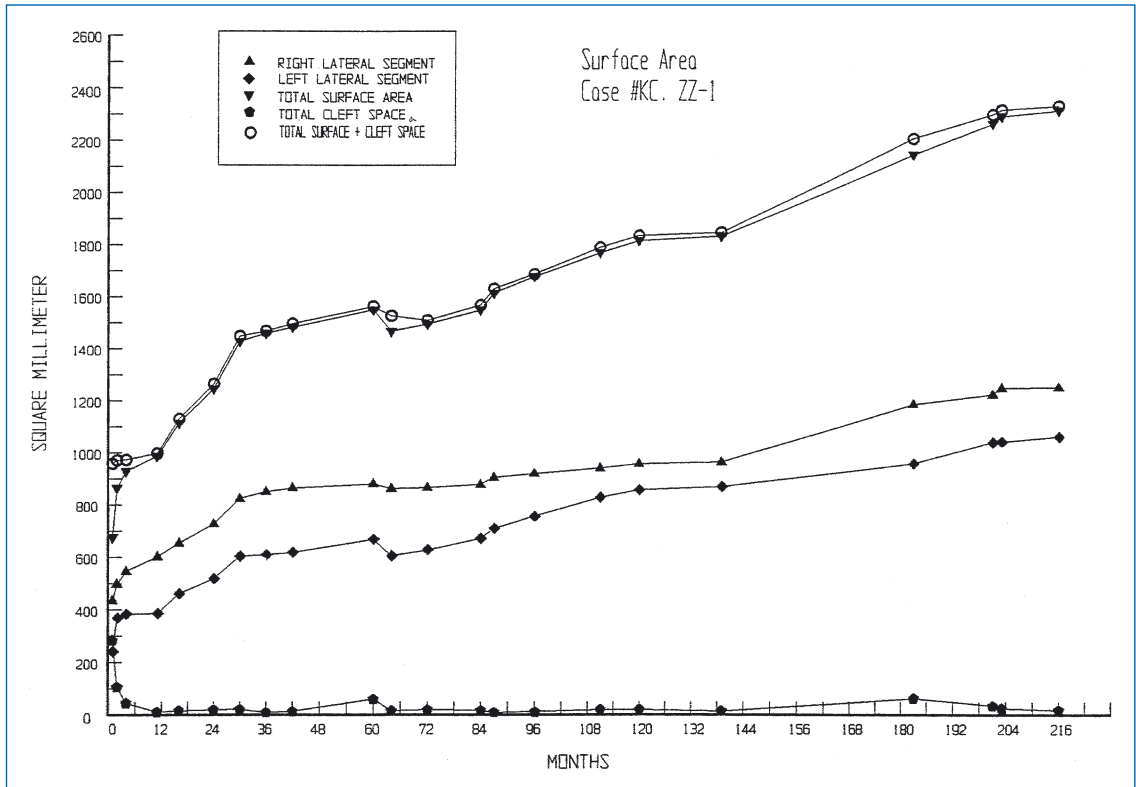


Fig. 6B.8. Case KC (ZZ-1). The palatal growth chart shows: (1) rapid growth acceleration in the first year which continues only slightly decreased until 36 months; (2) the palatal growth rate did not diminish after palatal surgery at 15 months; palatal growth slowed between 60 and 84 months, and then steadily increased; between 60 and 120 months, the growth of the lesser

cleft segment increased more rapidly than the nonleft segment; and (5) the palatal growth rate accelerated after 136 months. Comment: Based on palatal growth acceleration rates and the developing occlusion, one can safely conclude that palatal surgery did not interfere with its growth and development

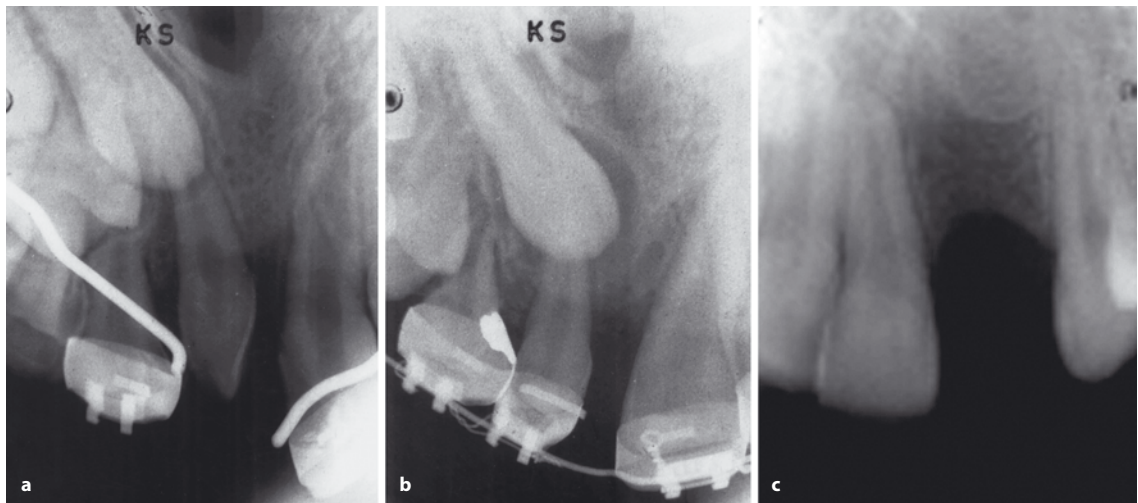


Fig. 6B.9 a–c. Case KC (ZZ-1). Tooth eruption into a secondary alveolar cranial bone graft performed at 7 years and 3 months of age. **a** The permanent lateral incisor is erupting into the graft. **b** Good root development, the lateral incisor is brought into the arch orthodontically. **c** Its root began to absorb and was extracted, good alveolar bone in the cleft space

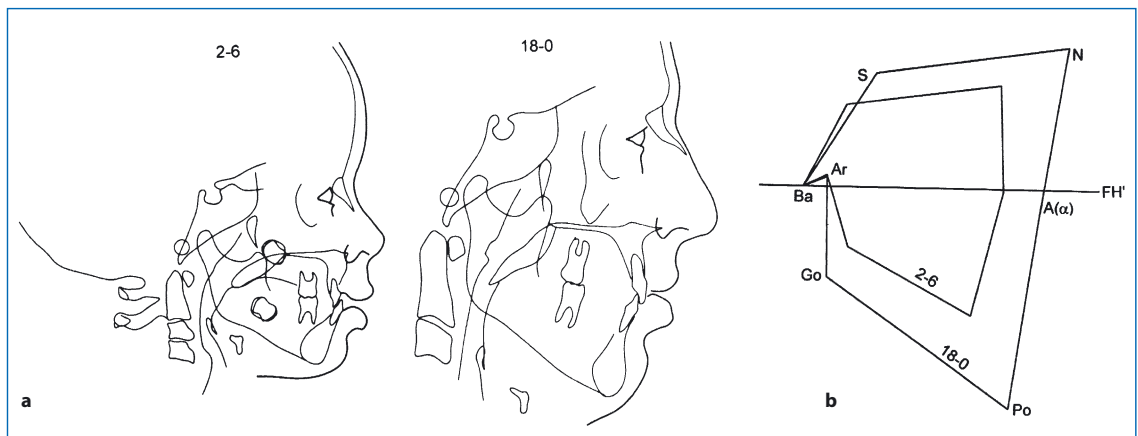


Fig. 6B.10. Case KC (ZZ-1). **a** Lateral cephalometric tracings and **b** superimposed polygons using Basion Horizontal Method (Coben). Both show an excellent facial growth pattern. At 18-0 the midface is slightly recessive but still very acceptable aesthetically

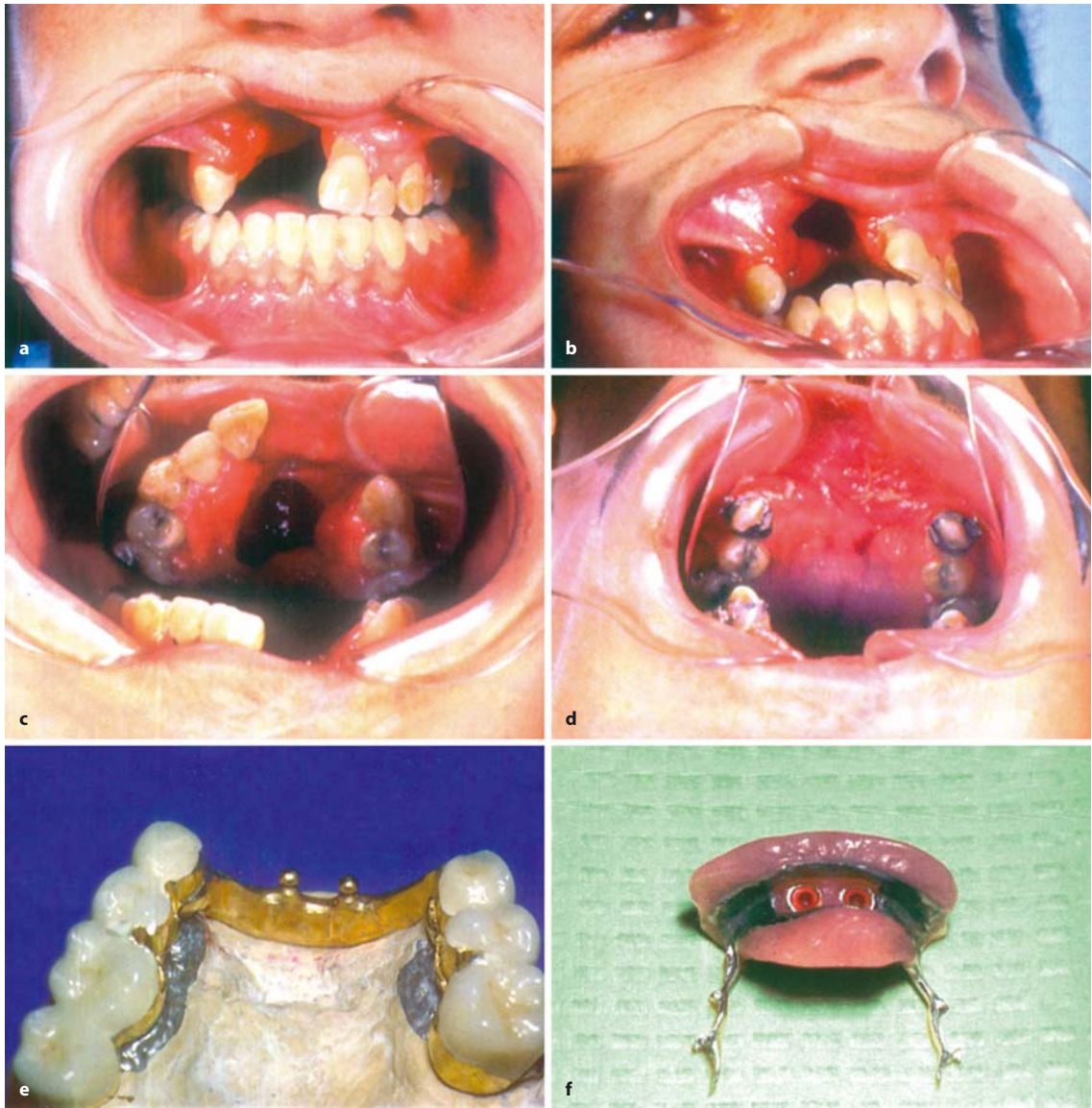


Fig. 6B.11 a-m. Case SP. Surgical and prosthetic treatment to replace a missing portion of a premaxillary segment and to close an oronasal opening (a transfer patient). Loss of blood supply to the right premaxillary area led to its exfoliation. Treatment plan: because the remaining blood supply to the left maxillary and central incisor was questionable and the teeth showed marked root absorption, the dentist (Alan Stoler) recommended their removal. The remaining teeth were to be crowned to support an anterior cast gold section to which a

removable prosthetic appliance would replace the missing incisor teeth and bumper the lip. **a, b,** and **c** Frontal and palatal view of an oronasal opening due to the loss of a portion of the right premaxillary segment. **d** Palatal view following soft tissue closure of the oronasal opening. **e** and **f** Anterior prosthetic appliance with splinted posterior teeth; anterior appliance with two holes and "o" rings to receive the two extensions on the anterior splint



Fig. 6B.11 a-m. (continued) **g** Posterior teeth splints with the anterior removable prosthesis in place on a model. **h** and **i** The gold anterior section spans the inter cuspid space. **j** Palatal view with the anterior prosthetic appliance in place. **k**, **l**, and **m**. Facial photographs showing good upper lip support with excellent dental aesthetics



Fig. 6B.12 a-k. Case JK (AF-64). Excellent facial and palatal growth in CUCLP when the palatal segments did not make contact after the lip was united. The lateral incisor is in position in the alveolar cleft area. **a-i** Serial facial and intraoral photographs show changes to the lip and nose after lip adhesion and

definitive lip surgery using Millard's Rotation Advancement procedure. Left facial asymmetry is apparent in the frontal photograph and is more noticeable in the intraoral photograph at completion of orthodontic treatment. The left side was kept in Class II occlusion



Fig. 6B.12 a-k. (continued) **j** and **k** Periapical films show bone closure of the alveolar cleft space with good lateral incisor alignment

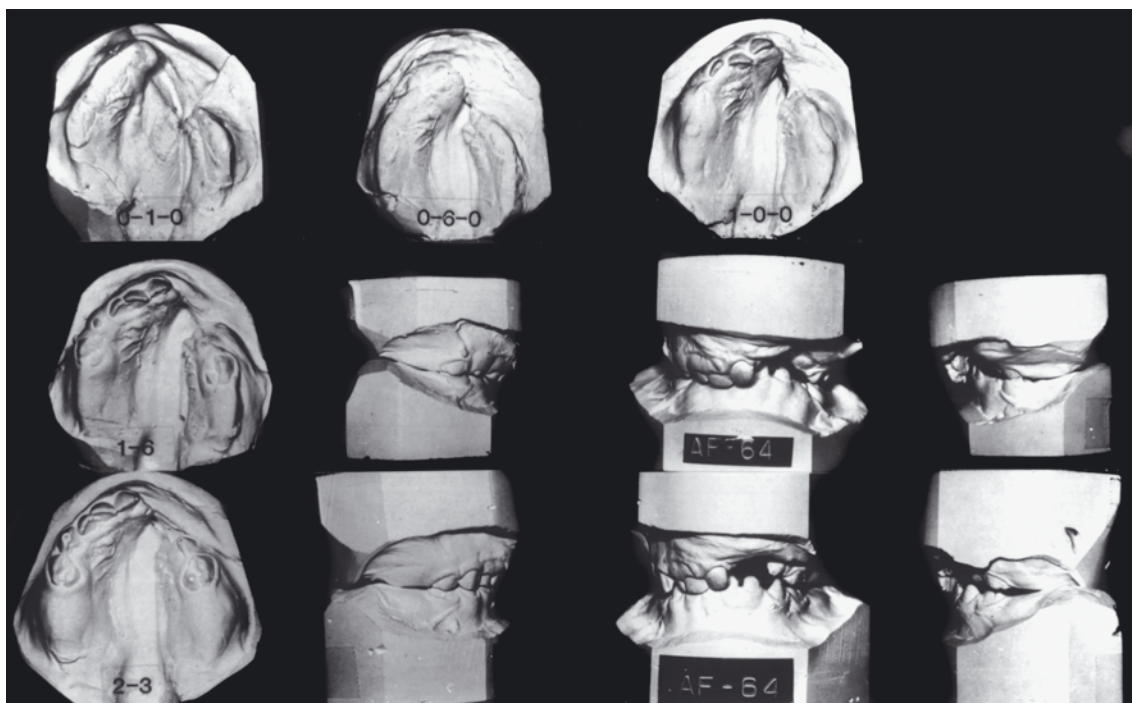


Fig. 6B.13. Serial casts of Case JK. Newborn: The nasal septum bows toward the cleft segment creating a very small cleft space. The great distance between alveolar segments is due to the upward tilt of the larger segment coupled with a small cleft segment. *0-6-0* After the lip is united, both palatal segments move toward the midline, narrowing the cleft space, more on the right

than the left side. However, the alveolar segments still do not meet due to the inferior turbinate on the lesser segment making premature contact with the septum preventing the lesser palatal segment from further medial movement. Note that the premaxillary portion of the larger segment has not moved medioposteriorly. *1-0-0, 1-6, 2-3, 3-2*

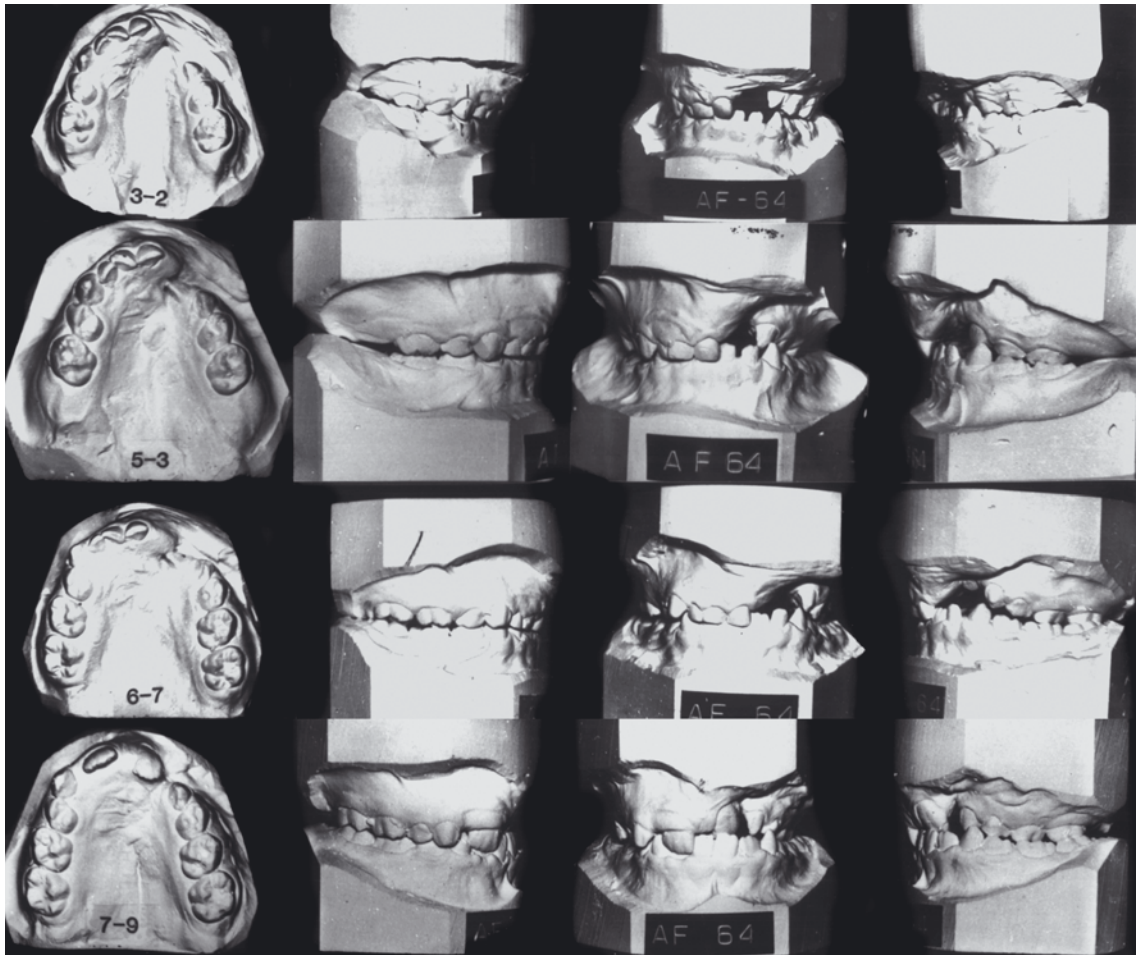


Fig. 6B.13. (continued) The palatal segments are still apart. 5-3. After removal of the inferior turbinate and with palatal closure, the tissue contracture created by the modified von Langenbeck procedure pulls the palatal segments together, placing the buc-

cal teeth in the cleft segment in crossbite. 6-7 The palatal segments have been expanded. 7-9 Without palatal arch retention the crossbite returned. The ectopically erupted left central incisor is in crossbite

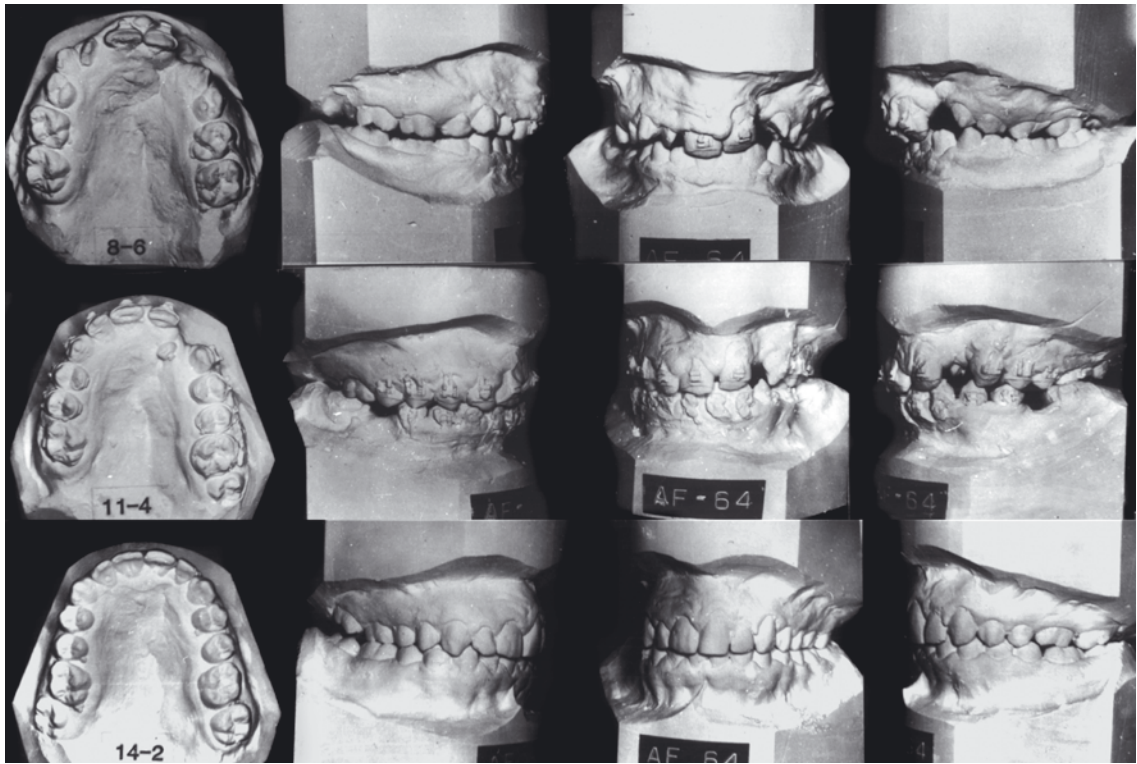


Fig. 6B.13. (continued) 8-6 Arch expansion mechanics were re-instituted and the left central incisor advanced into proper overjet. 11-4 and 14-2 Final orthodontic treatment was instituted and completed at 14 years of age. The impacted left lateral incisor was brought into alignment through the secondary alveolar cranial bone graft. Comment: After secondary alveolar bone grafting, arch expansion in most cases is stable. However,

in cases where new bone does not extend to the nasal aperture, we believe the buccal crossbite has a good chance of returning. The left side was in Class II occlusion, because it was not certain that the left lateral incisor could be properly aligned. If it was to be extracted, the cuspid would be positioned in the lateral incisor space

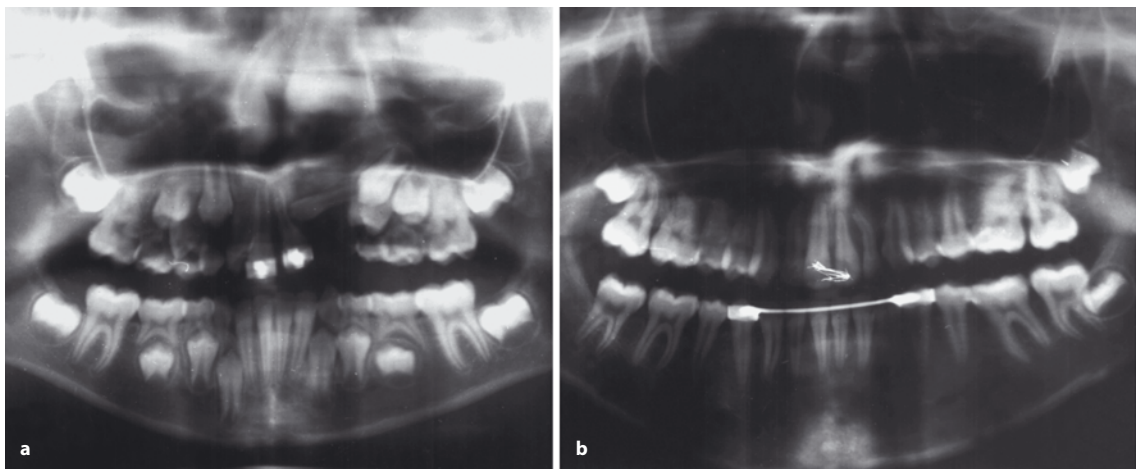


Fig. 6B.14 a,b. Case JK (AF-64). **a** Panorex: The left lateral incisor is palatally and horizontally impacted. **b** After treatment, the lateral incisor is well-aligned within the arch. Note that the

curvature to the root possibly occurred before it was fully formed and during orthodontic movement



Fig. 6B.15. Case JK (AF-64). Frontal cephaloradiograph shows that the nasal chamber on the cleft side is very narrow with a very flattened inferior conchae. The nasal septum is extremely bowed toward the cleft side. A lower cuspid to cuspid retainer is being worn

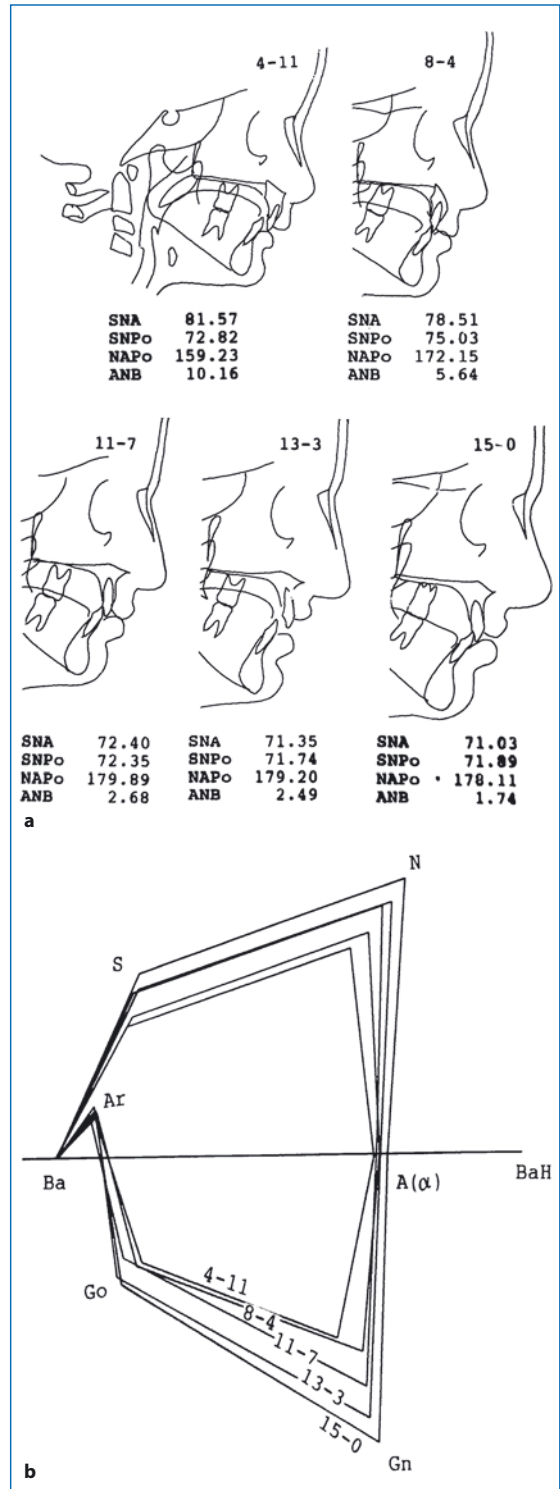


Fig. 6B.16 a,b. Case JK (AF-64). **a** Skeletal and soft tissue profile changes shown by lateral cephalometrics. The anterior projection of the midface and mandible relative to the anterior cranial base decreases with time as the profile flattens. The decreasing ANB angle reflects this change. **b** Superimposed polygons using the Basion-Horizontal method. This series clearly shows that the flattening of the skeletal facial profile occurred around 8 years of age and was brought about by the growth at the anterior cranial base and the mandible, whose plane angle increased with time. There was almost no forward growth of the midface between 4-11 and 13-3 years with only a small post-pubertal growth increment between 13-3 and 15-0 years of age



Fig. 6B.17 a–s. Case JD (AE 23). Complete unilateral cleft lip and palate. Excellent palatal and facial growth. A relatively large cleft space necessitated postponement of palatal closure until 20 months. Early secondary alveolar bone graft. Surgical history: Lip adhesion at 3 months followed by Rotation Advancement definitive lip repair at 6 months. Modified von Lan-

genbeck palatal cleft closure at 20 months. Secondary alveolar cranial bone grafts at 6 years. **a** Before and **b** after lip repair. **c** 2 years, 5 months. Anterior and buccal crossbite. **e** and **f** 3 years, 4 months. Anterior and buccal crossbite correction with fixed palatal expander

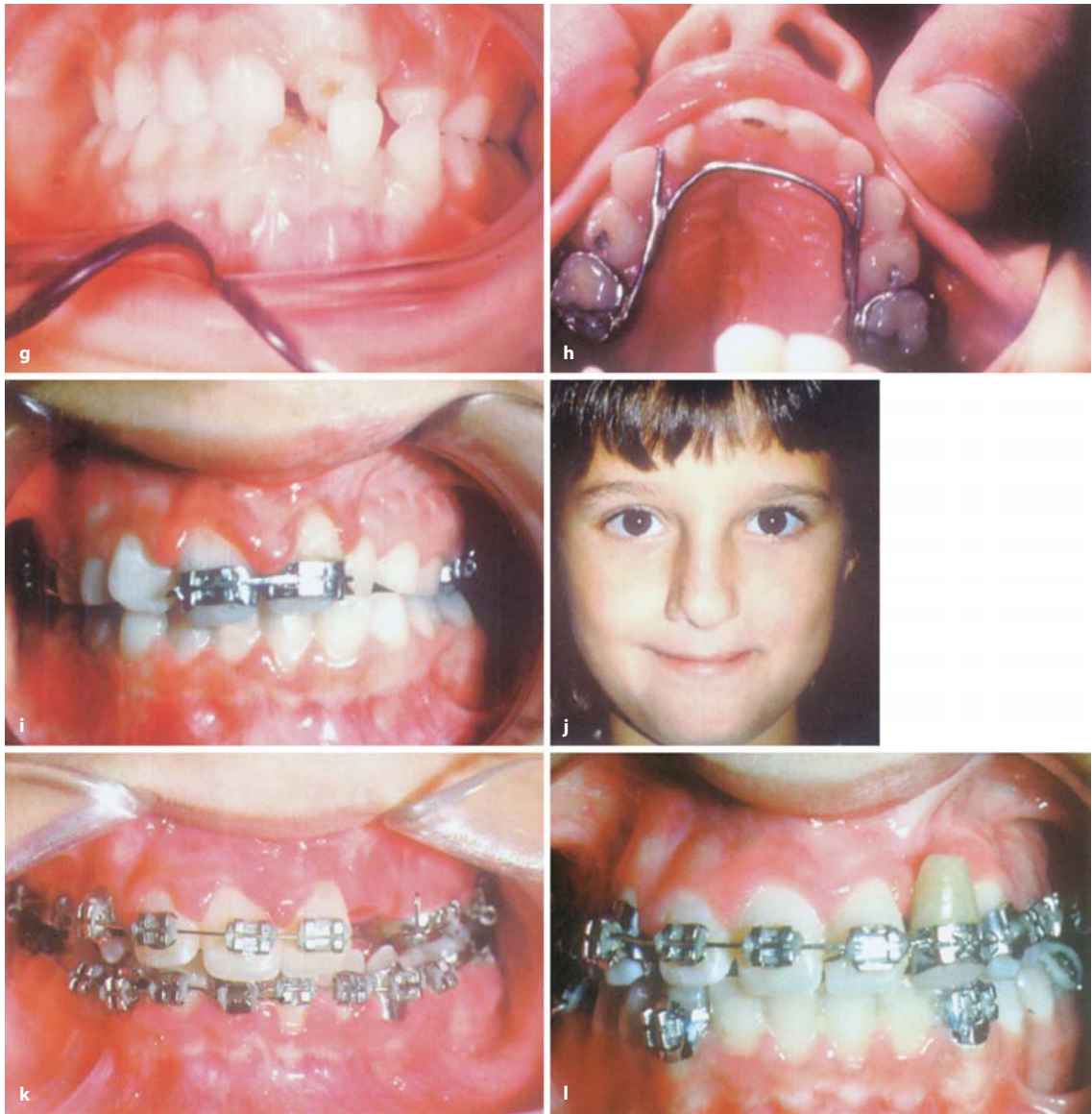


Fig. 6B.17 a-s. (continued) **g** and **h** 4 years, 6 months. After expansion, a fixed palatal retainer. **i** and **j** 9 years. Central incisor aligned in the mixed dentition. **k** and **l** Orthodontic appli-

ance with a false lateral incisor tooth with band attached to the orthodontic arch wire



Fig. 6B.17 a–s. (continued) **m–r** Facial and intraoral photographs at 15 years of age – on completion of orthodontic treatment. Ideal dental occlusion. **s** Maxillary retainer with an attached lateral incisor pontic tooth

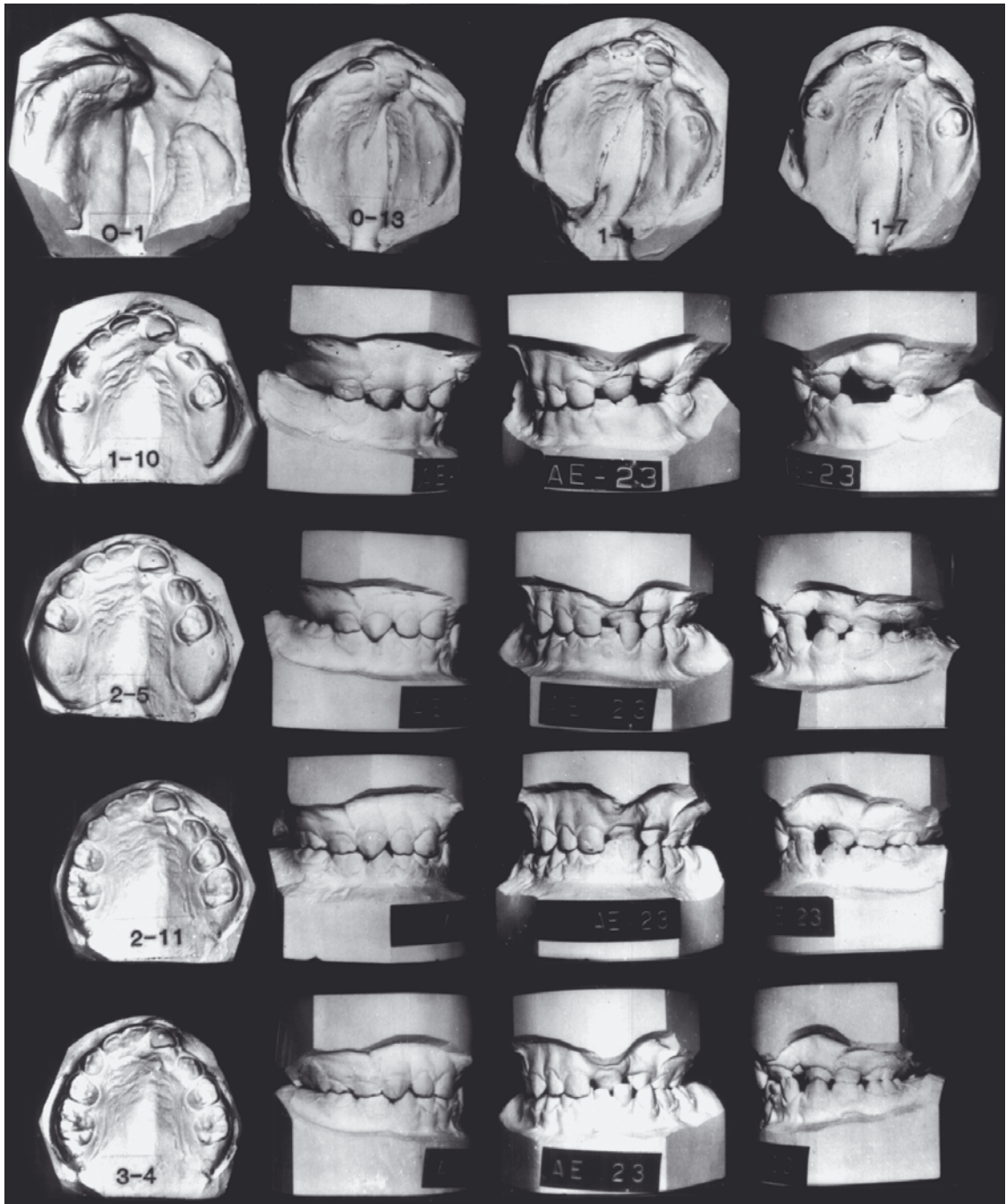


Fig. 6B.18. Serial casts of Case JD. 0-1 At birth. 0-13, 1-4, and 1-7 With the institution of compressive lip muscle forces by uniting the lip, the lesser cleft segment moved medially to make

contact with the vomer. The geometric changes to both segments brought the alveolar segments in good approximation. 1-10, 2-5, 2-11, and 3-4

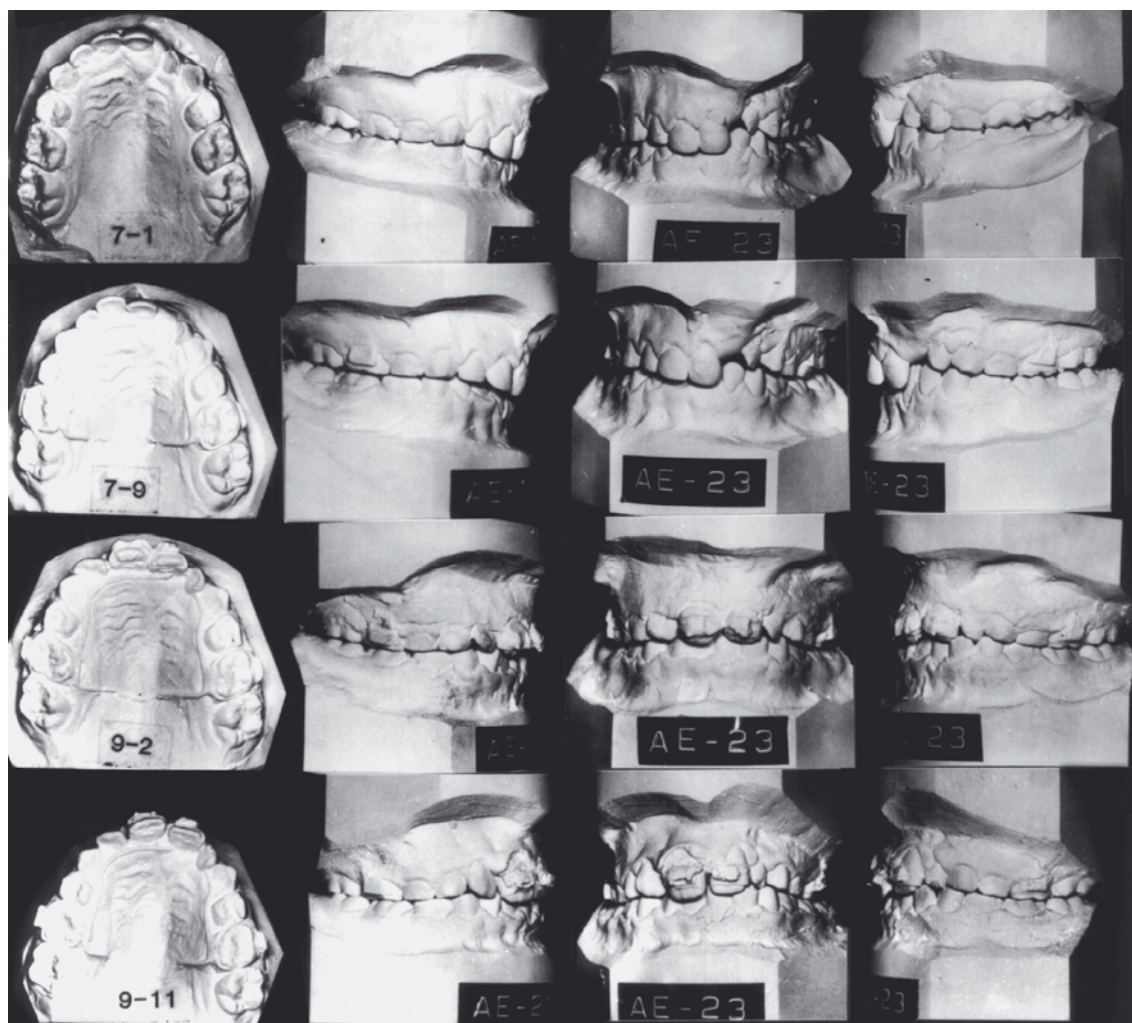


Fig. 6B.18. (continued) However, after palatal cleft closure at 20 months, the lesser segment moved further medially placing the left cleft segment in crossbite. Due to ectopic eruption, the left central incisor was in crossbite. After palatal expansion

and advancement of the left central incisor, excellent buccal occlusion was established. 7-1, 7-9, 9-2, and 9-11. Fixed palatal retainer is worn to maintain the arch form

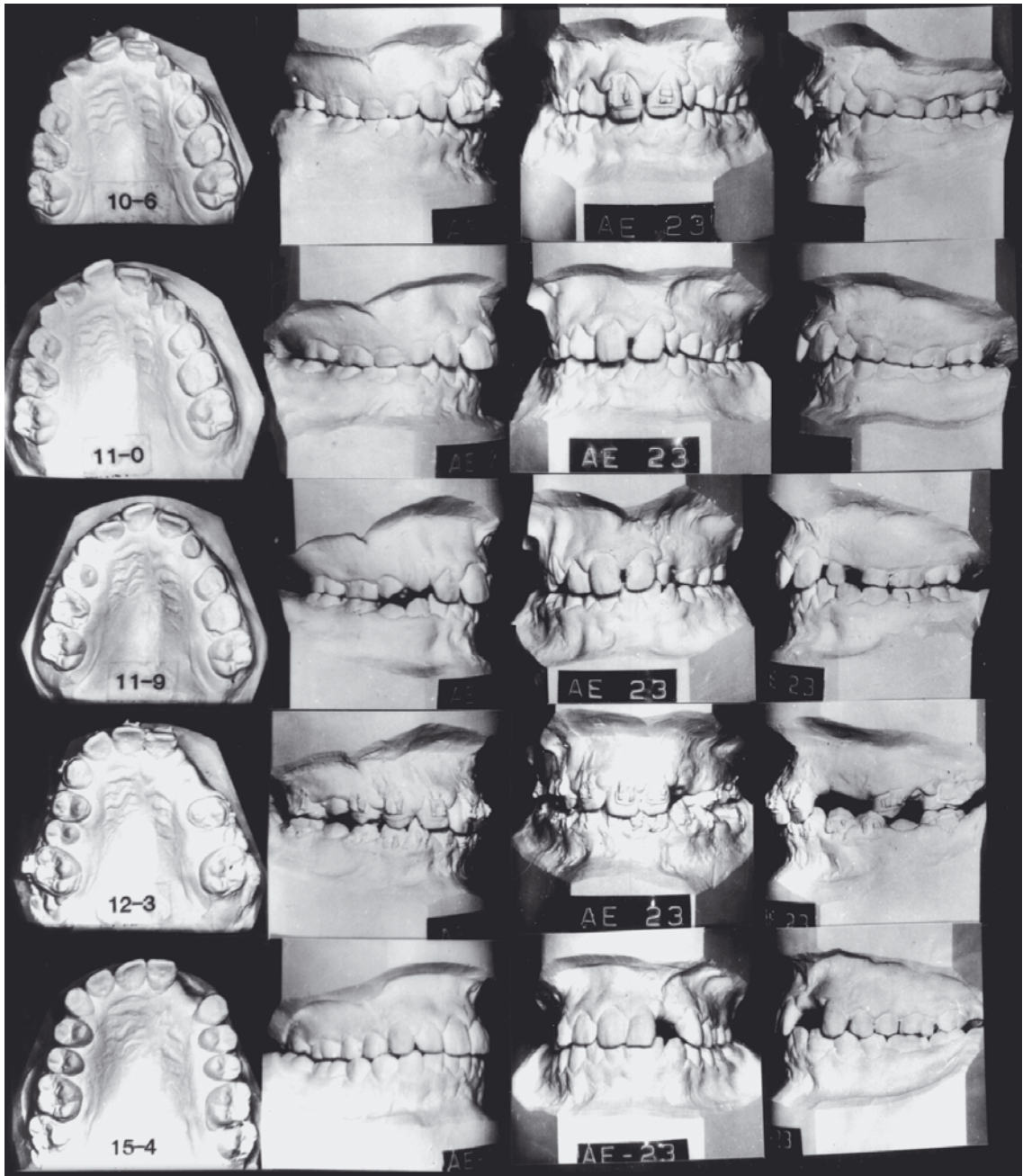


Fig. 6B.18. (continued) 10-6, 11-0, and 11-9. Palatal form retained with palatal appliance. The upper central incisors were rotated for aesthetic purposes. The left malformed lateral incisor was left in place until orthodontic treatment was instituted at 12 years of age. 15-4 Orthodontic treatment completed.

Note the slight flaring of the upper incisors and upper left cuspid, which was due to slight anterior maxillary bone deficiency. This is not an uncommon finding in complete unilateral clefts of the lip and palate.

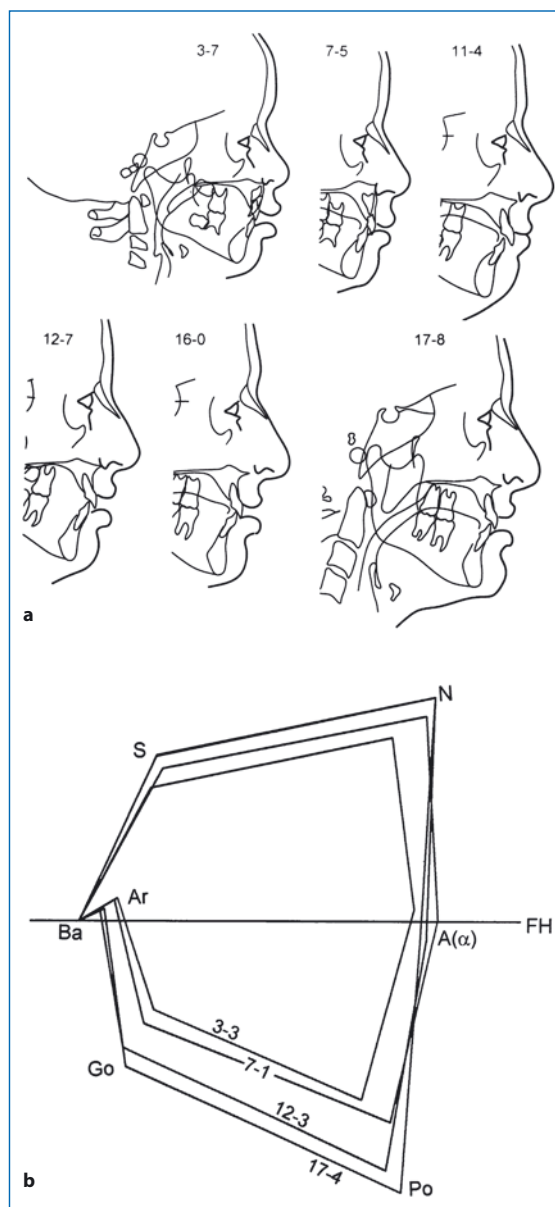


Fig. 6B.19. **a** Skeletal and soft tissue changes in Case JD. **b** Superimposed polygons using Basion Horizontal method (Coben). Both of these analyses show excellent facial changes. The midfacial protrusion actually reduced between 7-1 and 17-4. Comments: One of the main controlling factors in the treatment of children with clefts that involve the anterior bony segment is the amount of osteogenic deficiency in the area. In many noncleft children, some advancement of the anterior teeth is essential; however, advancing the anterior teeth in the child with a cleft results in flared incisor teeth because the bone deficiency prevents the roots from being brought forward, even with anterior root torque using rectangular arch wires

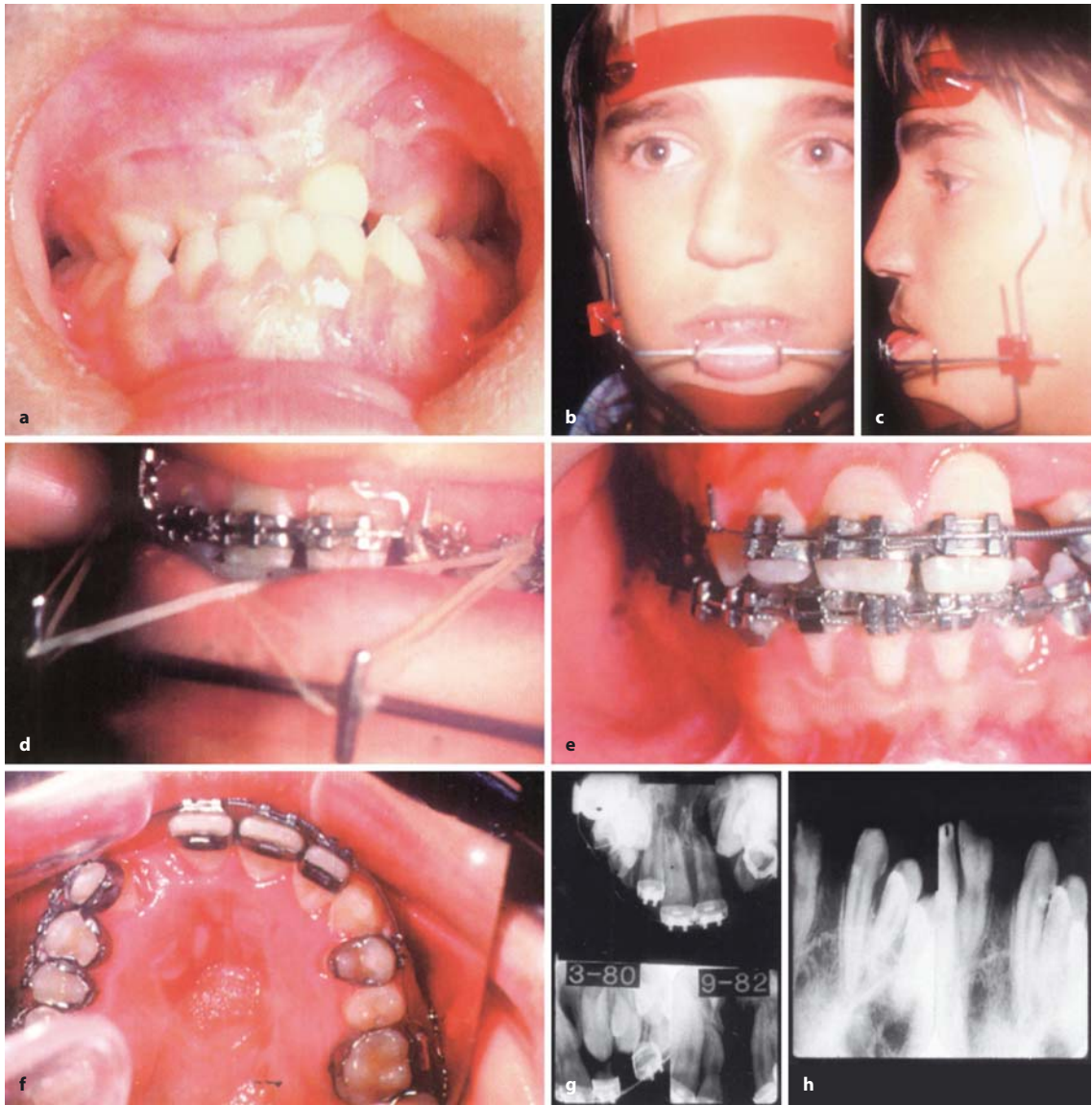


Fig. 6B.20 a–n. Case AB (EE-49). UCLP illustrating use of protraction maxillary orthopedics to correct midfacial retrusiveness secondary to growth-inhibiting scar tissue and/or maxillary osteogenic deficiency. Surgical history: Lip closure at 6 months. Hard and soft palate cleft closure at 16 months using an Island flap push-back. Secondary alveolar cranial bone graft at 10 years of age. **a** 2 years 11 months of age. Anterior and bilateral buccal crossbite could not be corrected in the deciduous or

mixed dentition. **b, c, d** Orthodontic-orthopedic forces to correct an anterior crossbite were initiated at 12 years of age using a Delaire-style protraction facial mask. **e, f** Ideal Class I (neutroclusion) with an ideal overjet and overbite. Palatal view shows thick transpalatal scar tissue caused by the Island flap. **g, h** Periapical films after secondary alveolar cranial bone graft. No left lateral incisor is present, but good cleft space closure is evident

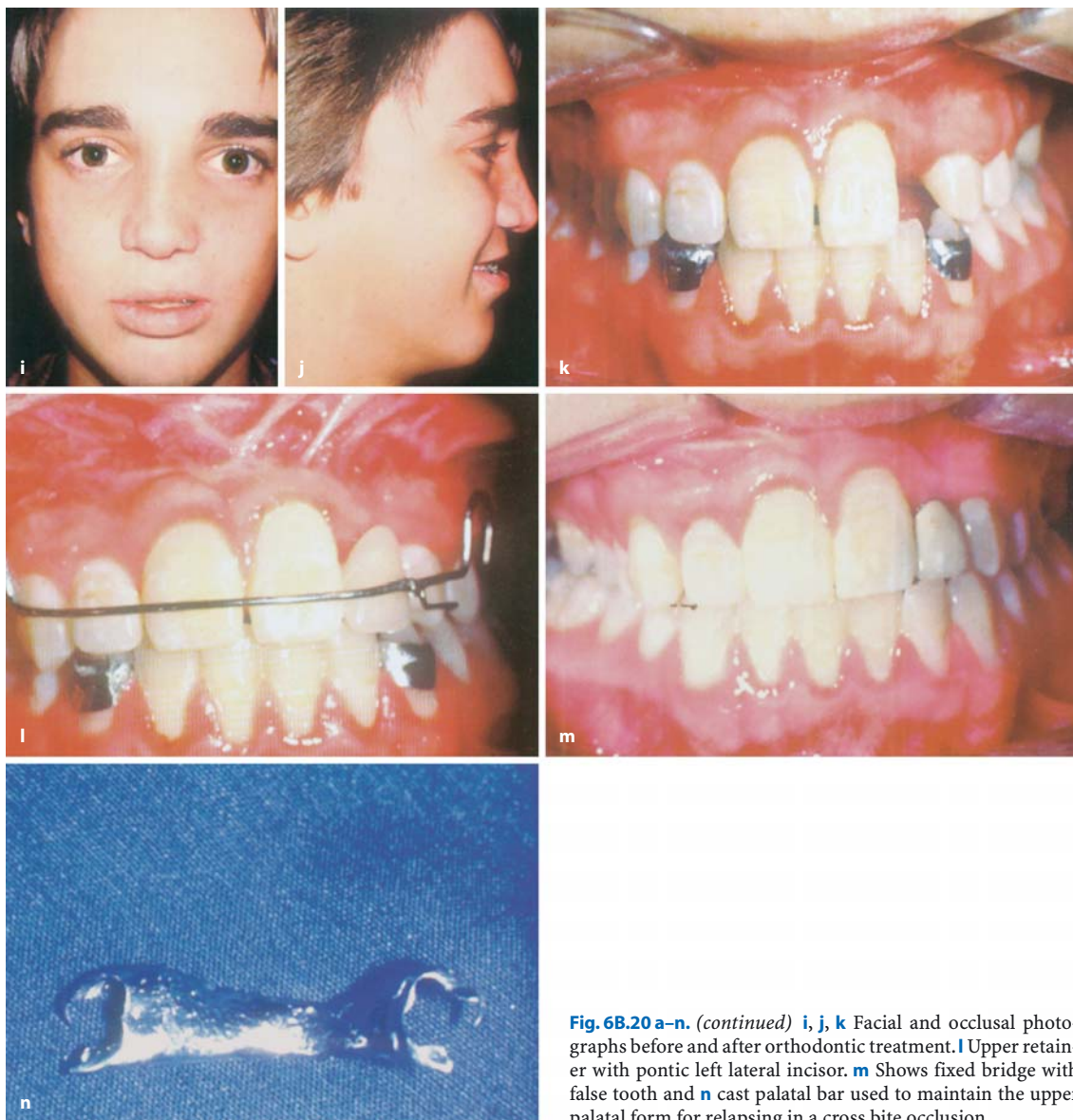


Fig. 6B.20 a-n. (continued) **i, j, k** Facial and occlusal photographs before and after orthodontic treatment. **l** Upper retainer with pontic left lateral incisor. **m** Shows fixed bridge with false tooth and **n** cast palatal bar used to maintain the upper palatal form for relapsing in a cross bite occlusion

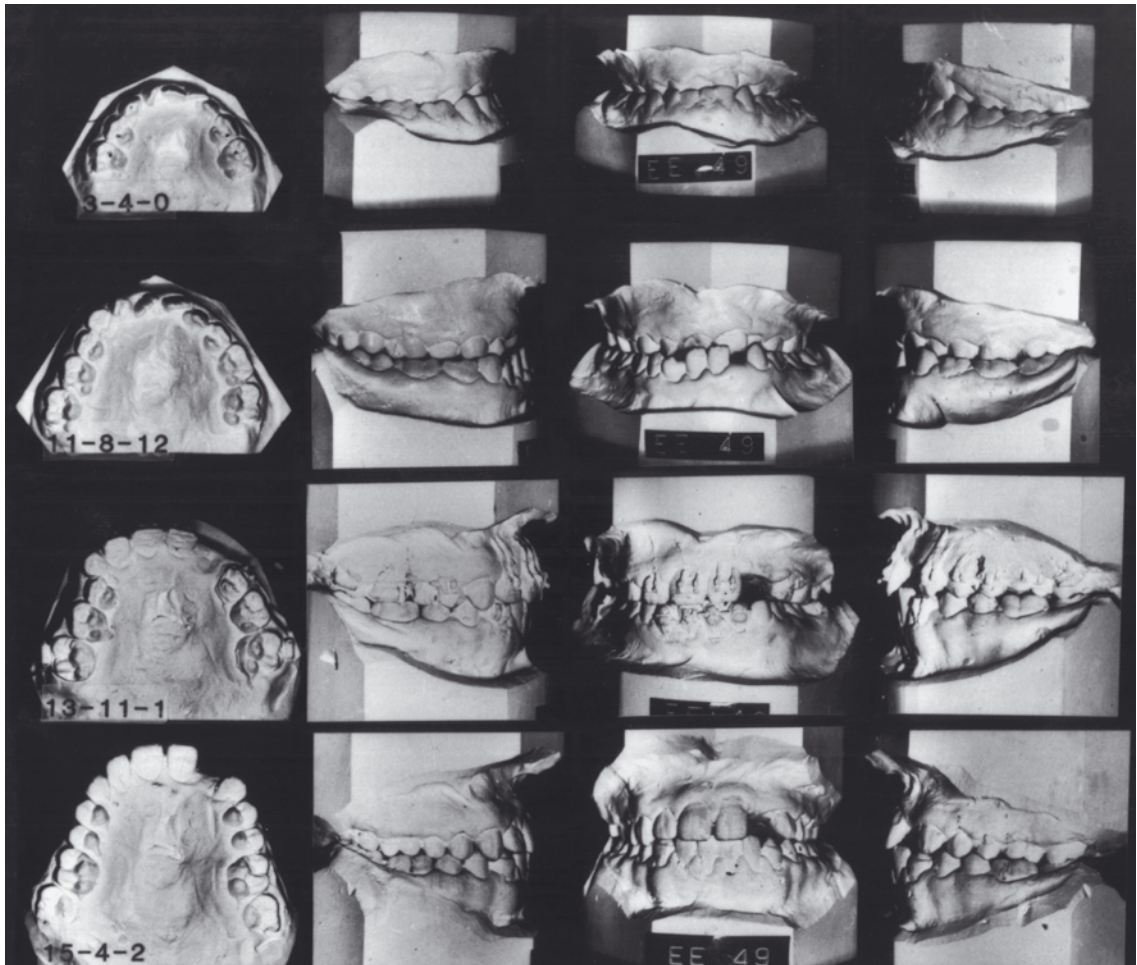


Fig. 6B.21. Serial casts of Case AB. 3-4-0 After Island flap hard and soft palate closure at 16 months of age resulting in bilateral buccal and anterior crossbites. 11-8-12 Occlusion just prior to orthodontic treatment. 13-11-1 After protraction mechanics using a Delaire style facial mask. 15-4-2 Occlusion after ortho-

dontics. Comments: Because maxillary deficiency is almost always present "A" point (subnasal) in the premaxillary area needs to be brought forward by using labile root torque on a rectangular arch

Fig. 6B.22 a,b. Case AB. **a** Cephalometric tracings at 14-11 and 16-2. **b** Superimposed polygons using Basion Horizontal method. A slight change in midfacial protrusion is noted after protraction forces were used to correct the midfacial retrusion and anterior crossbite. In this case the changes in the maxillary incisor axial inclination aided anterior crossbite correction more than maxillary protraction

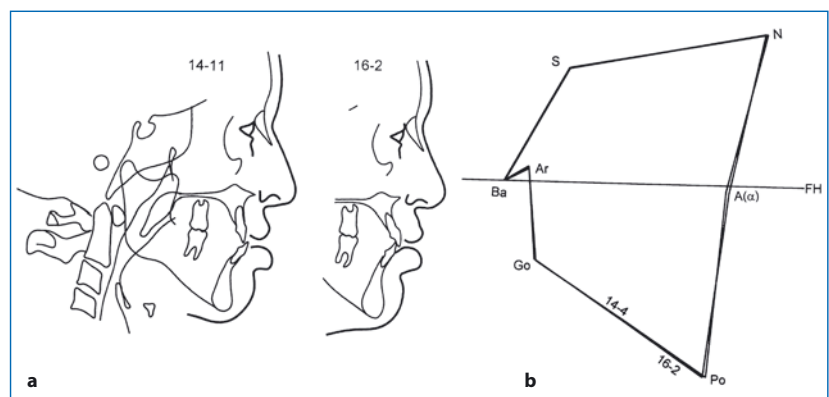




Fig. 6B.23. Conservative Treatment of a Patient with CUCLP. Lip adhesion at 3 months. Rotation advancement at 6 months. Palatal closure at 22 months using a von Langenbeck and vomer flap. Excellent occlusion and a flat face profile resulted

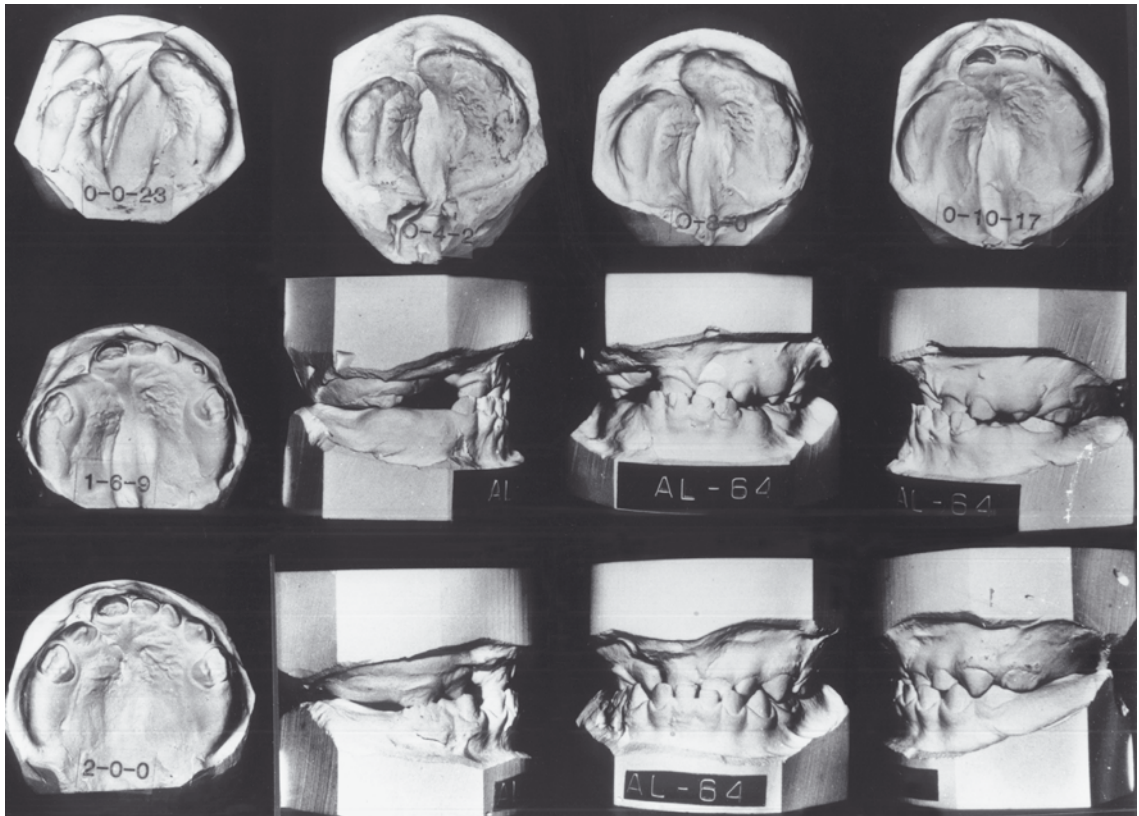


Fig. 6B.24. Case AL 64 Complete unilateral cleft lip and palate. Serial Casts: 0-0-23 to 2-0-0: Lip adhesion brings the overexpanded palatal segments together. The premaxillary portion of

the larger noncleft segments palatally positioned placing the teeth into an anterior cross-bite. The palatal cleft was closed at 1-11 with von Langenbeck plus vomer flap



Fig. 6B.24. (continued) 8-9-13 to 15-4-7: The maxillary anterior teeth were advanced into a proper overjet and overbite. Due to arch crowding the first bicusps were extracted and spaces closed. The alveolar bone graft at approximately 8 years permit-

ted the impacted lateral incisors to erupt into place. Note: the right lateral incisor crown is malformed but the root size and shape is normal. The crown will eventually be capped

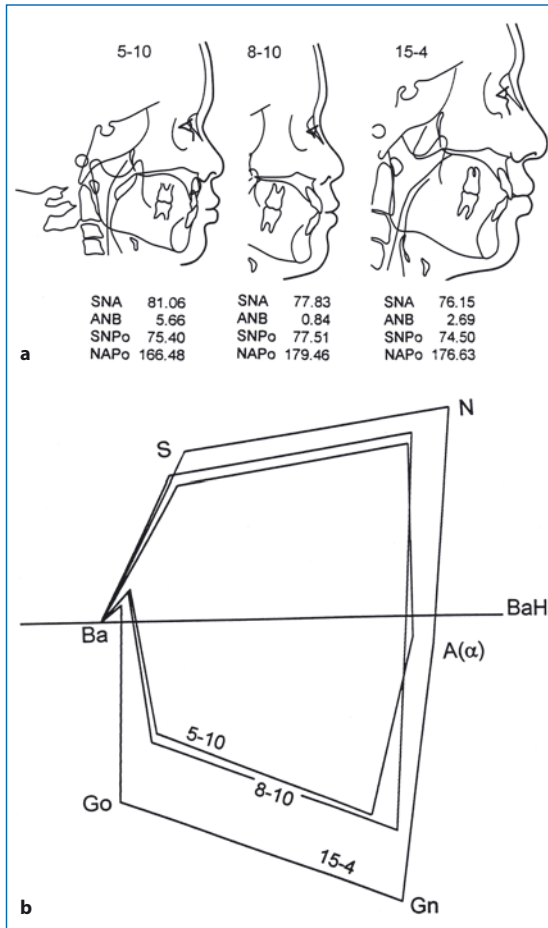


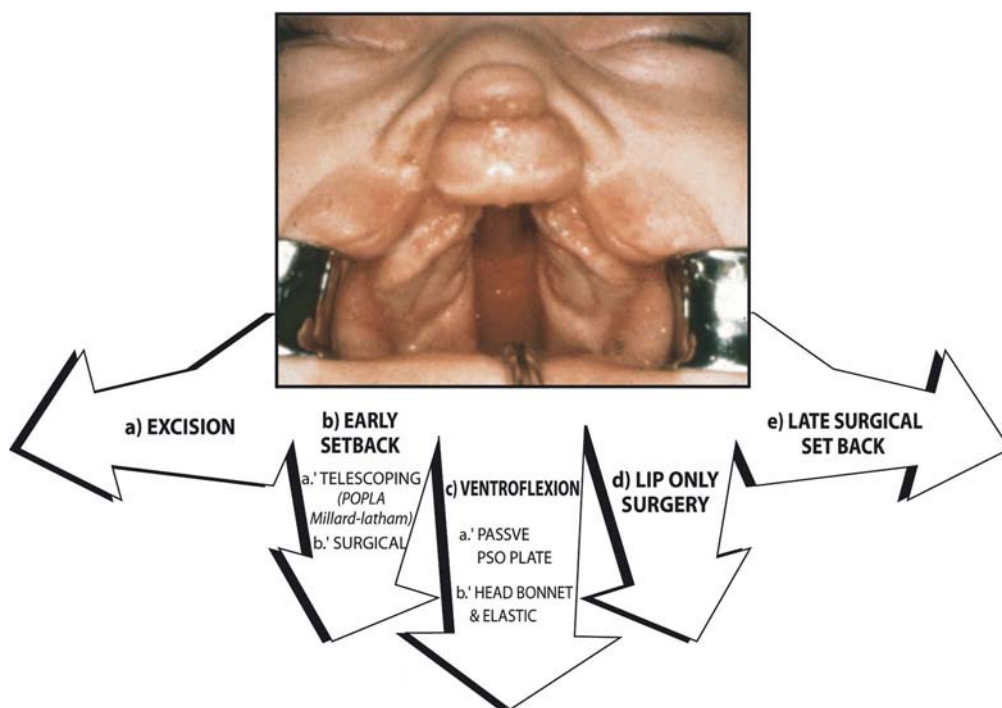
Fig. 6B.25 a,b. Case DM-AL 64. Very good facial growth pattern. The flattening of facial profile was dependent on good growth of all parts of the facial skeleton. The extraction of all the maxillary and mandibular first bicusps was necessary to retract the incisors and uncrowd the dentition

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6C Complete Bilateral Cleft Lip and Palate

SAMUEL BERKOWITZ



The surgeon is confronted with the following options when faced with a protruding premaxilla at birth:

1. Uniting the lip over the protruding premaxilla and considering later surgical setback and other surgical options.
2. External elastics attached to a head bonnet or elastic tape to the cheeks to ventroflex the premaxilla.
3. Early surgical premaxillary setback.
4. Complete removal (excision of the premaxilla).
5. Early mechanical retrusion prior to lip surgery or presurgical orthopedic treatment (PSOT) without retraction and with or without primary bone grafting or periosteoplasty.
6. Lip adhesion followed by definitive lip surgery at a later age.

6C.1 Premaxillary Protrusion: Real or Apparent. Is the Palate Deficient in Bone?

A bilateral cleft of the lip and palate can be complete or incomplete on one or both sides (see Chap. 6). Any number of variations can exist. In both incomplete

and complete bilateral clefts of the lip and alveolus, the size and shape of the premaxilla is dependent on the number of tooth buds and their distribution, making it symmetrical or asymmetrical (Fig. 6C.1). Because clefts of the lip/alveolus and the hard and soft palate come from different embryological sources, the cleft may involve the lip and alveolus with or without involving the hard and soft palate.

Based on the observations of Veau [1, 2] and Browne [3] and the later work of Friede and Pruzansky [4], Bergland and Borchgrevink [5], Harvold [6], Berkowitz [7], Friede [8, 9], Atherton [10, 11], and Handelsman and Pruzansky [12] the cause of premaxillary protrusion is the result of tension and the resulting bony overgrowth produced at the premaxillary vomerine suture (PVS) by displacement of that bone by the aberrant muscular forces of the detached outer musculature combined with the pushing force of the tongue fitting within the cleft (Fig. 6C.2). These investigators, using cephalometric data, concluded that the premaxilla is postured forward in the facial profile at birth (Fig. 6C.3).

Bergland and Borchgrevink [5] reported that, in complete bilateral clefts of the lip and alveolus with



Fig. 6C.1. **a** Small premaxilla: three deciduous incisors with one unerupted permanent central incisor. **b** Large premaxilla: four deciduous incisors with two permanent unerupted incisors. **c** Occlusal radiographs of premaxillae showing variations

in the number of anterior deciduous teeth and permanent tooth buds. **d** Occlusal radiographs of premaxillae showing variations in the number of anterior deciduous teeth and permanent tooth buds

Fig. 6C.2. **a** Line drawing of a complete bilateral cleft lip and palate with an arrow pointing to the premaxillary vomerine suture (PVS) **b** A small protruding premaxilla IBCLP extends forward of the facial profile and lateral palatal segments. **c** Variations in bilateral cleft lip and palate. The size of the premaxilla varies with the number of teeth it contains. Classification is dependent on the completeness of clefting of the lip and alveolus and whether there is a cleft of the hard and soft palate. Yet one or both sides of the hard palate may or may not be attached to the vomer. If it is attached to the vomer, it is classified as being incomplete. Even in complete clefts of the lip and alveolus the extent of premaxillary protrusion will vary. **A** Incomplete bilateral cleft lip and palate. Complete cleft lip and palate – left side.

Incomplete cleft lip and palate – right side. **B** Complete bilateral cleft lip and palate. Complete cleft palate – both sides. **C** Incomplete bilateral cleft lip and palate. Incomplete palatal clefts – both sides. **D** Complete bilateral cleft lip and palate. Incomplete right and complete left palate. **E** Incomplete bilateral cleft lip and palate. Incomplete left palate and complete right palate. **F** Complete bilateral cleft of the lip and palate. Incomplete right and left palatal segments. **G** Complete bilateral cleft of lip and palate. Incomplete left palate and complete right palatal segment. **H** Complete bilateral cleft lip and palate. Incomplete left palate and complete right palate. **I** Incomplete bilateral cleft lip and alveolus. Normal palate

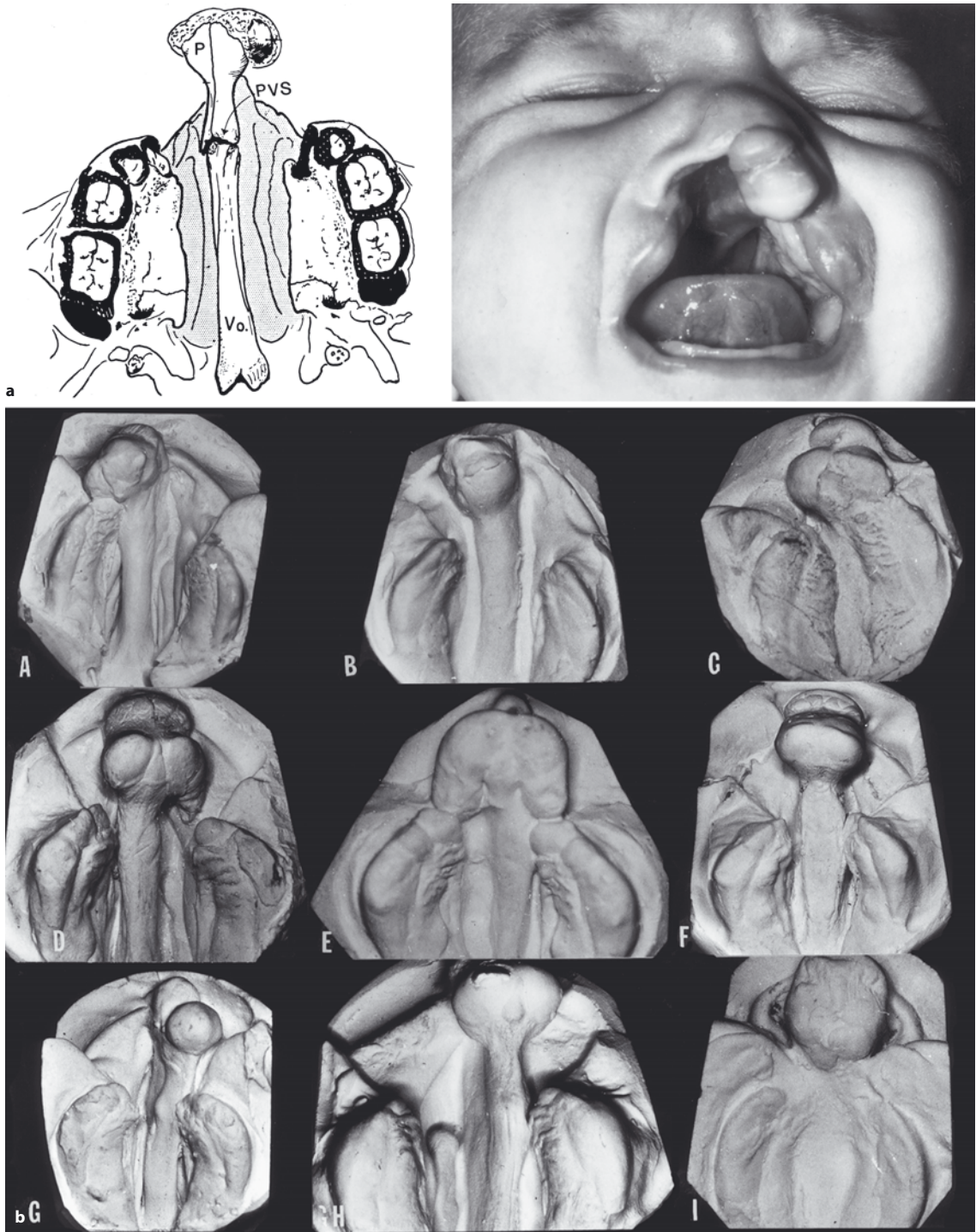


Fig. 6C.2 a, b.

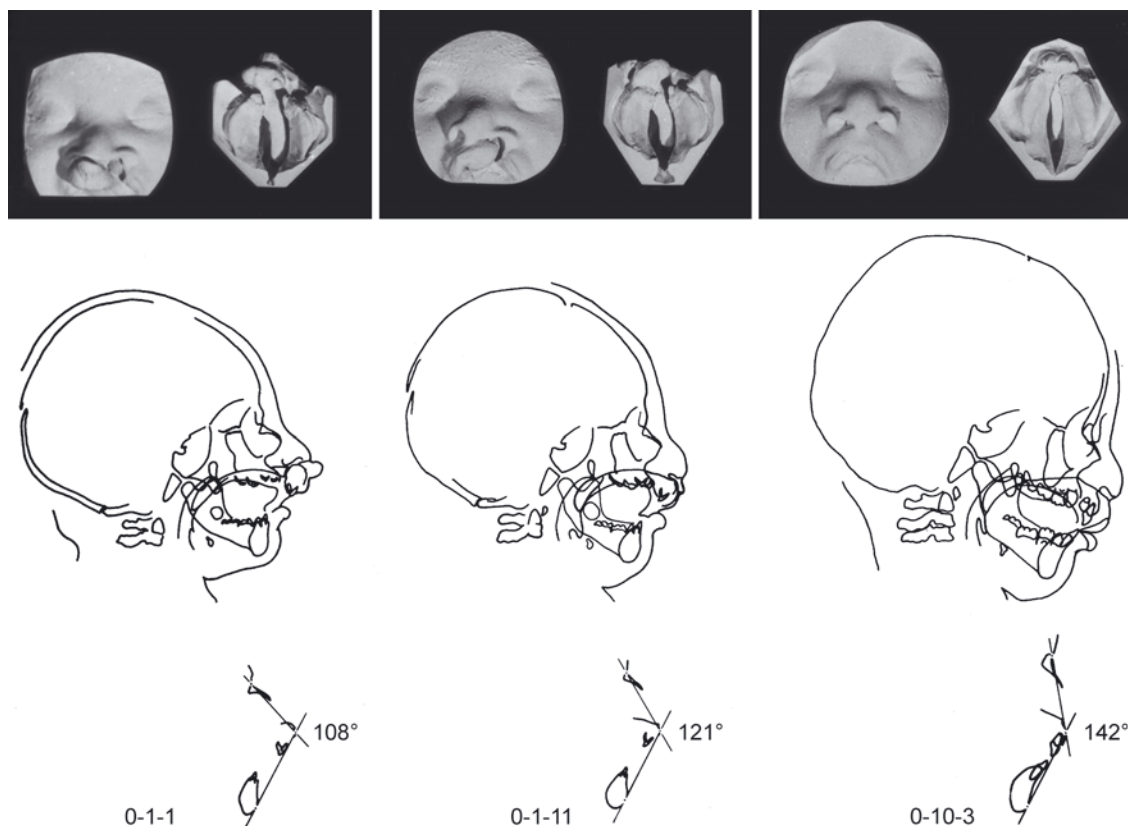


Fig. 6C.3. Two-stage lip closure in a CBCLP. Ten months after lip closure the angle of facial convexity changed from 108° to 142° as a result of the ventroflexion of the premaxilla at the pre-

maxillary vomerine suture (PVS) coupled with some additional mandibular growth. A two-stage lip closure is rarely necessary. (courtesy of S Pruzansky)

intact palates, the premaxilla was protrusive but palatal size was well within normal limits. In these cases, the septum was detached from the normally developed palates, and the protrusion of the premaxilla was interpreted as representing a premature release of the normal growth potential of the septum. The premaxilla apparently reached its geometric position within the skull at an earlier time prior to birth, yet the palatal segments followed a normal growth rate. Berkowitz's clinical records support the findings of Coup and Subtelny [13], who reported marked palatal hypoplasia in bilateral clefts of the lip and palate. Berkowitz reports in this book that palatal growth rates are highly variable according to the amount of scarring created when closing the cleft space.

6C.2 The Premaxillary-Vomerine Suture

Pruzansky [14, 15] and Friede and Morgan [16] used metal implants on either side of the premaxillary-vomerine junction to demonstrate cephalometrically that this suture was a major site of bony overgrowth (Fig. 6C.4). Earlier, Berkowitz [7] and later Pruzansky [15], Friede and Morgan [16], Pruzansky and Friede [17], Friede [18], Vargervik [19], and Berkowitz [20] suggested that the overgrowth was probably a secondary reaction to the lack of restraint from the cleft orbicularis oris muscle. They showed that midfacial growth continued for 1–3 years even after the lip was united but at a slower rate. Berkowitz's [7] serial cephalometric tracings and digital cast study demonstrated that the midface continued to protrude for 2–3 years after the lip was united, then its forward growth slowed markedly (Fig. 6C.5). Not surprisingly, in some instances, the facial convexity did not change due to the continued forward growth of the maxilla and premaxilla but was due to more vertical rather

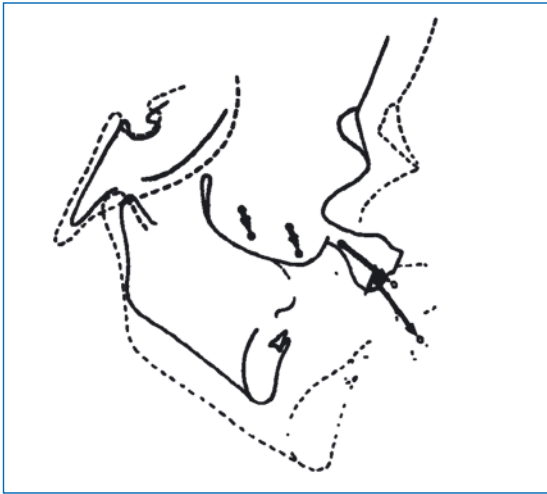


Fig. 6C.4. To test growth at the premaxillar vomerine suture (PVS) in complete bilateral cleft of the lip and palate, two pairs of metal pellets were placed on either side of the PVS at 6 months of age. At 3 years, 5 months of age, the distance between the anterior and posterior sets of pellets had increased with the growth at PVS. Note that there was no change in distance within each set of metal pellets (courtesy of S Pruzansky)

than forward growth of the mandible, maintaining the facial convexity (Figs. 6C.6, 6C.7).

Pruzansky [14, 15], Friede [18], and Atherton [10, 11] described the premaxillary vomerine suture as resembling other facial sutures. Friede and Morgan [16] confirmed the presence of small islands of cartilage in

the suture; these were secondary occurrences resulting from mechanical stresses and not part of a force system causing growth.

Burston [21, 22] and Latham [23, 24] following Scott's [25, 26] "Nasal Septum Growth" thesis, believe that the displacement is not real but only apparent. Burston thought that the lateral segments of the maxilla were retroplaced and considered the premaxilla to be in normal position. Latham suggested that a contributing factor in producing the projecting premaxilla was the shortening of the septomaxillary ligament which drew it forward.

Pruzansky and Friede's [17] roentgencephalometric data confirmed the existence of a true, rather than a relative, premaxillary protrusion. Metallic implants were inserted on either side of the PVS at the time of initial surgery and were followed up roentgencephalometrically. Although continuous growth was recorded in the PVS, there was a postsurgical decrease in the premaxillary protrusion. On the basis of both the histology and cephalometrics, Friede [18] concluded that traumatic surgery involving the PVS would be likely to contribute to the impaired midfacial growth. This has been verified by Berkowitz's more recent follow-up studies of the effects of mechanical premaxillary retraction on profile development. He has observed similar midfacial problems of impaired growth (see Chap. 20).

Berkowitz speculated that severe pressure at the PVS would cause a hemorrhage followed by fibrosis and synostosis with cellular destruction, both acting to inhibit sutural growth. In contrast, a united lip

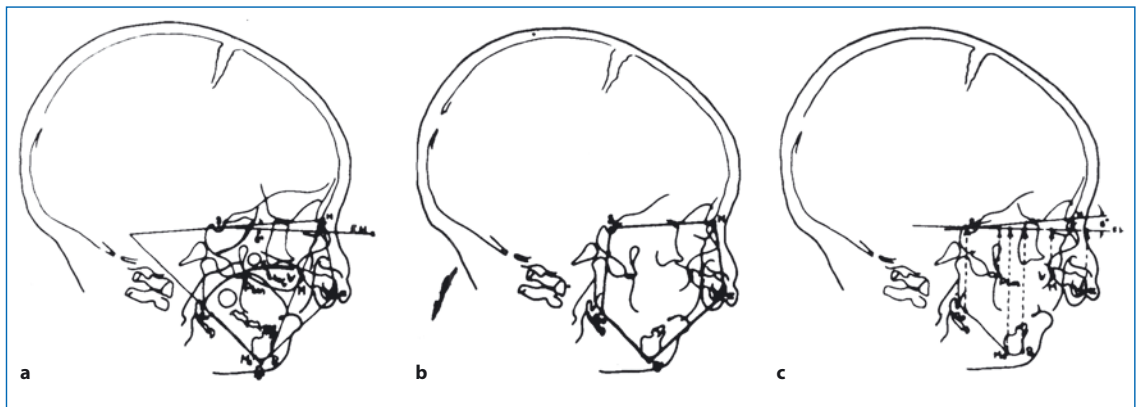


Fig. 6C.5 a-f. A method used to show serial changes in facial growth and development in CBCLP using Sella for registration while superimposing on the anterior cranial base. Facial angles and landmarks (upper left): Nasion (N), Pogonion (Po), Gonion (Go), Sella (S), Alpha (a) most anterior point on the premaxilla, Menton (Me), constructed Frankfort horizontal (FHc), Anterior point of the lateral palatal processes (M), Pterygomaxillary fissure (Ptm). Facial polygon (upper middle) drawn connecting

landmarks S-N-a-Po-Me-Go-S. Landmark points (upper right) projected to a constructed Frankfort horizontal line which is drawn 6° from the anterior cranial base (SN) at S. **b** An example of excellent facial growth. The cultural standard for a good aesthetic Caucasian face is a "flat" face, having an angle of facial convexity of approximately 180°. Most newborn noncleft faces have a relatively acute facial profile associated with relative retrognathia which usually flattens with growth

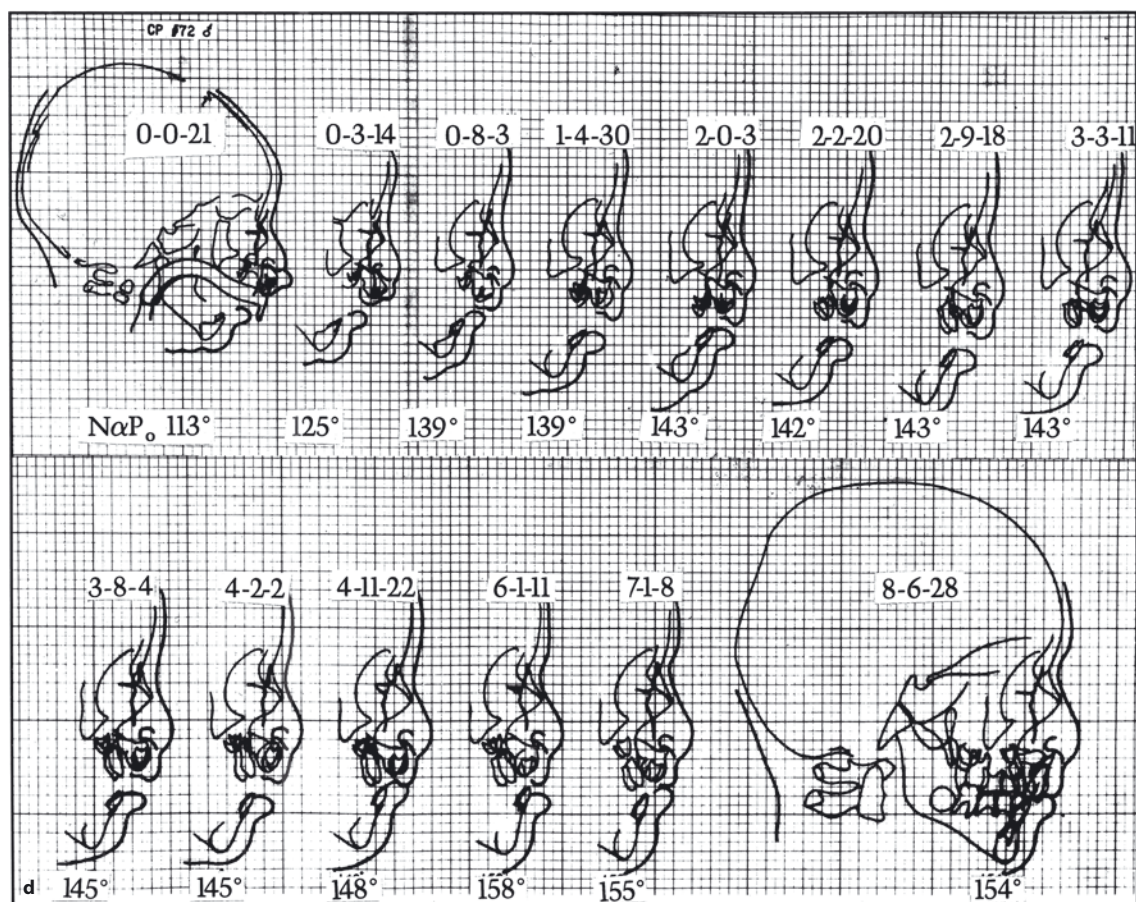


Fig. 6C.5 a-f. (continued) d Serial lateral cephalometric tracings showing changes in the angle of facial convexity (NaPo) from 113° to 154° in 8 11/42 years.)

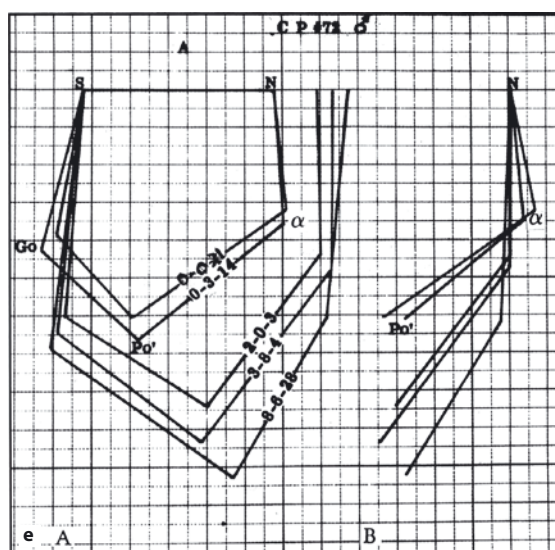


Fig. 6C.5 a-f. (continued) e Facial polygon of the case shown in Fig. 6C.5d. Each polygon showing profile changes is superimposed on SN and registered at Sella. The midfacial protrusion at (a) had not increased after 2 years, whereas both the anterior cranial base (N) and mandible had increased in size. The mandible grew forward and downward, flattening the facial profile. The timing of these growth changes is variable

across the protruding premaxilla exerts a more tolerant retrusive force within the PVS, acting well within its physiologic threshold level for slowing down bone growth but not totally inhibiting it.

Based on conclusions drawn from his clinical findings and those of Pruzansky and Friede's [17] research, Berkowitz rejects Latham's [23, 24] "force of the cartilaginous septum via the septo-premaxillary ligament" as the cause of premaxillary protrusion. Enlow's [27] description of the nasomaxillary complex's growth is worth repeating:

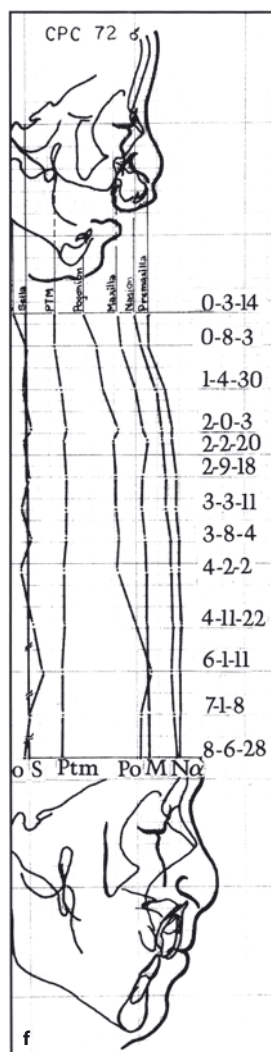


Fig. 6C.5 a-f. (continued)

f Projection of landmark points to the constructed Frankfort horizontal line drawn 6° from SN at S. This growth projection system also shows that the midfacial protrusion did not increase after 2 years, whereas the forward projection of the mandibular body increased markedly until 8 11/42 years of age. These are the main factors leading to the flattening of the facial profile. Good facial growth changes can occur early or late (from [70])

The nasomaxillary complex is in contact with the floor of the cranium. The whole maxillary region in total is displaced downward and forward away from the cranium by the expansive growth of the soft tissues in the midfacial region. This then triggers new bone growth at the various sutural contact surfaces between the nasomaxillary composite and the cranial floor. Displacement thus proceeds downward and forward as growth by bone deposition simultaneously takes place in an opposite upward and backward direction.

McNeil [30] mistakenly had stated that the detached palatal segments from the nasal septum are not only reduced in mass but also are not brought forward with the developing nasal septum.

This failure of the palatal segments to move forward with growth would lead to a retrusive midface

with a Class III malocclusion (he did not want to blame the suggestions).

6C.3 Facial Growth Studies Show That Midfacial Retrusion Is Not Predictable

Semb [28] and Ross [29] established that, in the cleft population, both the maxilla and mandible – not solely the maxilla – are repositioned within the face. If McNeil's [30] belief that the bilateral cleft palatal segments are left behind in their growth was accurate, a greater proportion of the cases must show a Class III malocclusion of one or both sides. As previously mentioned, Berkowitz's [31] mixed cross-sectional study of the occlusion in CBCLP determined that the maxillary complex was not positioned posteriorly relative to the mandible. Also, a buccal crossbite is not a predictable outcome, as McNeil had suggested. Semb [28] and Ross [29] concluded that, although the midface and mandible are positioned posteriorly within the face, the maxilla is not repositioned relative to the mandible and that McNeil's hypothesis that the maxilla in complete clefts of the lip and palate is retruded and needs to be brought forward was in error.

6C.4 Long-Term Facial Growth Findings Show Class III Outcomes Are Not Predictable

Semb [25] conducted a serial lateral and frontal cephalometric study of 90 cases from the Oslo archives with bilateral cleft lip and palate. Since 1962, the treatment procedure has involved uniting the lip and closing the hard palate cleft space in two stages. No presurgical orthodontics were utilized because the surgeon and Bergland, an orthodontist and director of the program, believed that any bilateral cleft lip can be closed without presurgical palatal manipulation. In the period spanning 1950 to 1960, a von Langenbeck procedure was performed to close the hard palate cleft between 3 and 4 years of age; after 1960, the timing of the closure was reduced to 18 months of age. Secondary alveolar bone grafting using cancellous iliac crest bone was performed prior to the eruption of the permanent canine teeth.

Twenty-five percent of the cases needed superior-based pharyngeal flaps, which were performed before the child started school. No orthodontics were utilized in the deciduous dentition. Protraction headgear was used in the mixed dentition in one third of the cases. Fixed retention was necessary in all cases. Semb's study showed that: (1) the maxilla progressively receded over time; (2) the mandible was retrusive

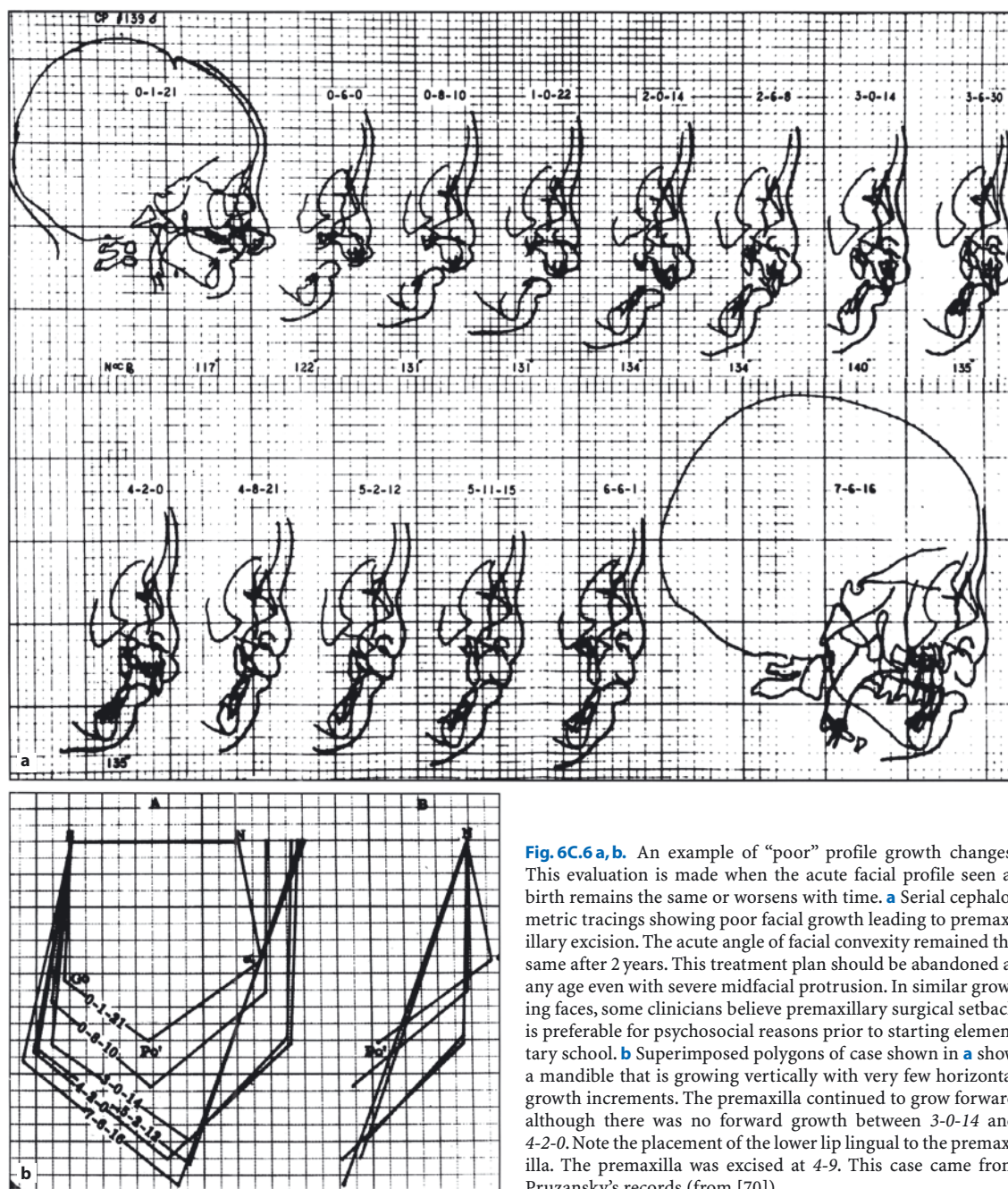


Fig. 6C.6 a, b. An example of “poor” profile growth changes. This evaluation is made when the acute facial profile seen at birth remains the same or worsens with time. **a** Serial cephalometric tracings showing poor facial growth leading to premaxillary excision. The acute angle of facial convexity remained the same after 2 years. This treatment plan should be abandoned at any age even with severe midfacial protrusion. In similar growing faces, some clinicians believe premaxillary surgical setback is preferable for psychosocial reasons prior to starting elementary school. **b** Superimposed polygons of case shown in **a** show a mandible that is growing vertically with very few horizontal growth increments. The premaxilla continued to grow forward although there was no forward growth between 3-0-14 and 4-2-0. Note the placement of the lower lip lingual to the premaxilla. The premaxilla was excised at 4-9. This case came from Pruzansky’s records (from [70])

with a steep mandibular plane with an increased gonial angle; (3) anterior lower face height was elongated and posterior facial height reduced; (4) the facial growth pattern was notably different from the normal Bolton standards: (a) male and female facial growth patterns were similar except that the males’ linear dimensions were larger; (b) the prominent premaxilla

would gradually realign in the preschool years; and (c) surgical premaxillary setback was never required. Berkowitz’s unpublished serial cast cephalometric data support these statements (unpublished data).

Vargervik’s [19] cross-sectional study of 51 males with BCLP treated with a variety of primary procedures (excluding premaxillary setback) showed pro-



Fig. 6C.7. The first 2 years of maxillary and mandibular tracings of the cases with good (see Fig. 6C.5) and bad (see Fig 6C.6) facial growth are compared to show the reasons for these evaluations. CP-72: Good facial growth. In this case, the degree of premaxillary protrusion relative to the lateral palatal segments (the anterior cleft space) was markedly reduced with growth. Premaxillary protrusion relative to the anterior cranial base was reduced as well. CP-139: Poor facial growth. The degree of premaxillary protrusion relative to the lateral palatal segments

remained the same, while the premaxillary protrusion relative to the anterior cranial base increased. (Courtesy of S Pruzansky). Comments: Although mandibular growth in the two cases was similar in degree, the superimposed polygons show that the vertical direction of growth of the “bad” grower’s mandible and not its size was the determining factor for the changes in the angle of facial convexity (*NaPo*). Tension at PVS created by the lower lip positioned lingual to the premaxilla increased premaxillary growth

file values similar to those reported by Hellquist et al. [32], Dahl [33], Smahel [34], Semb [28], and Friede and Johanson [35]. The Oslo team’s average for maxillary prominence and lower face height were slightly more favorable. Narula and Ross [36], reporting cross-sectional data on thirty 6-year-old subjects and mixed longitudinal data on 34 subjects with BCLP treated conventionally without surgical setback and vomer flap, concluded the maxillary length reached normal values at 16 years of age.

In the Swedish sample followed longitudinally by Hellquist et al. [32], similar facial convexity was also noted, although both the maxilla and mandible were reported to be slightly more prominent. The patients analyzed by Hellquist et al. [30] had a two-stage lip closure, pushback palatoplasty, and, at an average age of 6, a delayed periosteoplasty.

Friede and Johanson [35] reported facial growth in 13 Swedish children with bilateral clefts of the lip and palate (BCLP), five at age 7, and eight at age 10 years. The patients, who had had lip adhesion and vomer flap (without premaxillary setback) and velar closure with push back, also exhibited facial convexity similar to the Oslo sample.

Friede and Pruzansky’s [4, 5] cephalometric report of 27 North American children in three treatment groups who were followed to 17 years of age do, however, show some differences in comparison with the Oslo sample. Six subjects treated by early premaxillary setback and seven treated by late setback (3–8 years of age) had profile values similar to those reported by the Oslo group. However, 14 subjects with no premaxillary setback had average values significantly more convex than those reported for the Oslo and other samples. None of the North American sub-

jects had had vomer flaps, and it is implied that a more convex facial profile will be obtained if vomerplasty is excluded from primary surgery.

Also, the negative growth affect of the surgical premaxillary setback may become evident at a later age.

6C.5 The Vomer Flap: Good or Bad? Are all Vomer Flaps the Same?

Semb's [28] report on the longitudinal data of the Oslo group is critical of those who condemn the vomer flap. She states that the possible growth-retarding effect of a vomer flap has been discussed by several authors [4, 16, 35, 37, 39, 40–44]. Friede and Pruzansky [4] observed more favorable growth in patients treated without a vomer flap; however, this is not a uniform finding in the comparative studies. Some clinical centers not using a vomer flap have shown results similar to those where a vomer flap has been utilized. The extent to which the observation of a growth-retarding effect of a vomer flap can be generalized remains in some doubt, and may reflect other variations in sample composition or surgery.

The effects of a vomer flap on facial growth have also been considered by the Oslo CLP team [43] which reported the flap to be clinically insignificant. Only one patient in their sample of 90 cases exhibited any degree of maxillary retrusion where surgical maxillary advancement was judged necessary.

In the opinion of the Oslo team, a vomer flap provides the particular advantages of early separation of the nasal and oral cavities without artificial obturators, a low prevalence of symptomatic fistulae, an acceptable arch form, and a good foundation for mixed dentition alveolar bone grafting [46].

Berkowitz's finding of serial complete bilateral cleft lip show results similar to those of the Oslo group. A modified vomer flap was used in conjunction with a von Langenbeck procedure. He believes that the negative effect of a vomer flap on midfacial growth and arch form, as described by Prysdo et al. [47] has not been conclusively proven. Patients who have needed LeFort I advancement of only the lateral palatal segments have had very large anterior cleft spaces in the permanent dentition even after the posterior hard palate had been surgically closed at 18–24 months of age with a vomer flap. The premaxillae in these cases were in good overbite-overjet relationships prior to moving the palatal segments into a Class II relationship to close the missing lateral incisor(s) space. Other times the lateral incisor spaces are left open.

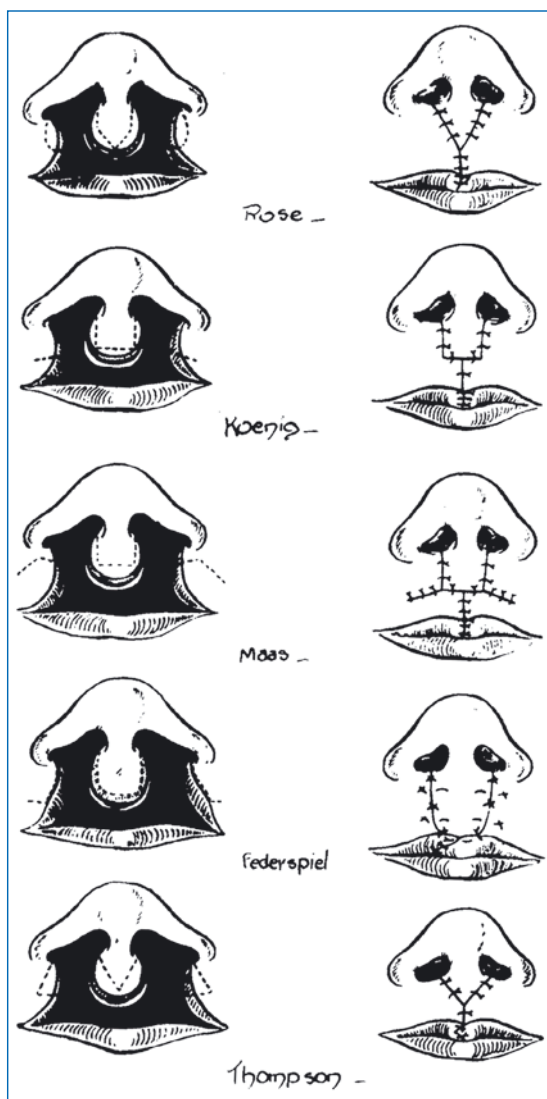


Fig. 6C.8. Various surgical techniques used to unite the lip in BCLP. Experience has shown that the best results are obtained when the prolabium is used to construct the entire midportion of the lip. (from [71])

6C.5.1 External Elastics Attached to a Head Bonnet or Elastic Tape Strapped to the Cheeks (Fig. 6C.9)

These force delivery systems will reduce premaxillary protrusion prior to lip surgery, and only need to be utilized for approximately 1 or 2 weeks. Elastic forces exert the same backward pressure against the protruding premaxilla as a lip adhesion. There are no valid reasons to object to the use of external facial elastic traction with arm restrains to prevent its removal forces for 1–2 weeks prior to lip surgery.



Fig. 6C.9 a–c. A head bonnet with an elastic strap (external facial traction) placed against premaxilla is an efficient and painless procedure to ventroflex the premaxilla prior to lip surgery. **a** At birth. **b** With head bonnet in place. **c** After lip repair.

The head bonnet aids the surgeon in reducing tension at the lip suture site. The force generated at PVS is less than that created by using mechanical premaxillary retraction (see Chap. 20)

6C.5.2 Uniting the Lip (Figs. 6C.8–6C.10)

In all complete clefts of the lip and palate at birth, the wide dislocation of the lateral palatal segments coupled with the protruding premaxilla can be gradually overcome by following a treatment protocol that allows the palatal segments to move into proper orientation by the application of the physiological forces of a united lip without the need for presurgical orthopedic treatment. Lip adhesion is followed by definitive lip surgery and observation of facial and palatal changes (Fig. 6C.8). If necessary, later surgical premaxillary setback, or surgically moving the lateral palatal segments forward to close a very large persistent anterior cleft space, can be considered.

Clinicians who favor this procedure believe the degree of midfacial protrusion will decrease with facial growth under the influence of the midfacial growth-restraining forces of the united lip [4, 7, 13, 14, 19, 48–51].

Findings by Bishara and Olin [50] and Aduss et al. [51] support the conclusions drawn from earlier facial growth studies by Berkowitz [7], Friede and Pruzansky [4], Handelman and Pruzansky [12], and Vargervik [19] that the protruding premaxilla tends to be molded back by lip pressure and can sometimes be aligned within the lateral palatal segments without resorting to neonatal maxillary orthopedics. If not, then the premaxilla in most cases can be aligned within the arch in the deciduous dentition using a fixed orthodontic quad helix appliance.

The use of lip adhesion within the first 3 months followed by a definitive lip revision, usually within the first 6 months of age, permits the surgeon to perform

surgery in stages as the face changes. Logan's Bow can be used to reduced the strain at the lip suture when the definitive lip surgery is performed. Surgeons who favor stage treatment procedure are willing to postpone obtaining a maximum early aesthetic result for what they believe will offer superior long-term benefits. The most severe, unaesthetic premaxillary overbite in the deciduous and mixed (transitional) dentition, even with a buccal crossbite, will not interfere with dental function or swallowing or have any long-term deleterious effects on speech and midfacial growth.

Serial facial photographs indicate that surgical procedures that use the entire prolabium for the central portion of the lip produce the best aesthetic results. A long, tight lip is the predictable result when the lateral lip elements are brought together beneath the prolabium (see Fig. 6C.10a, b).

6C.6 Profile Changes (Figs. 6C.14–6C.18)

Hanada and Krogman [52], Berkowitz [7], Bishara and Olin [50], Boyne [53], Vargervik [19], Handelman and Pruzansky [12], Friede [9], Semb [28], and Narula and Ross [38] all have performed longitudinal studies of the changing soft-tissue profile of bilateral cleft lip and/or palate (BCL/P) and reported that, with the slow resolution of the protruding premaxilla with growth, the profile became more harmonious in appearance. In the Oslo study of BCLP, Semb [25] concluded that early dentofacial orthopedics were not a necessary precursor to lip and palate closure in order to attain long-term positive profile changes.

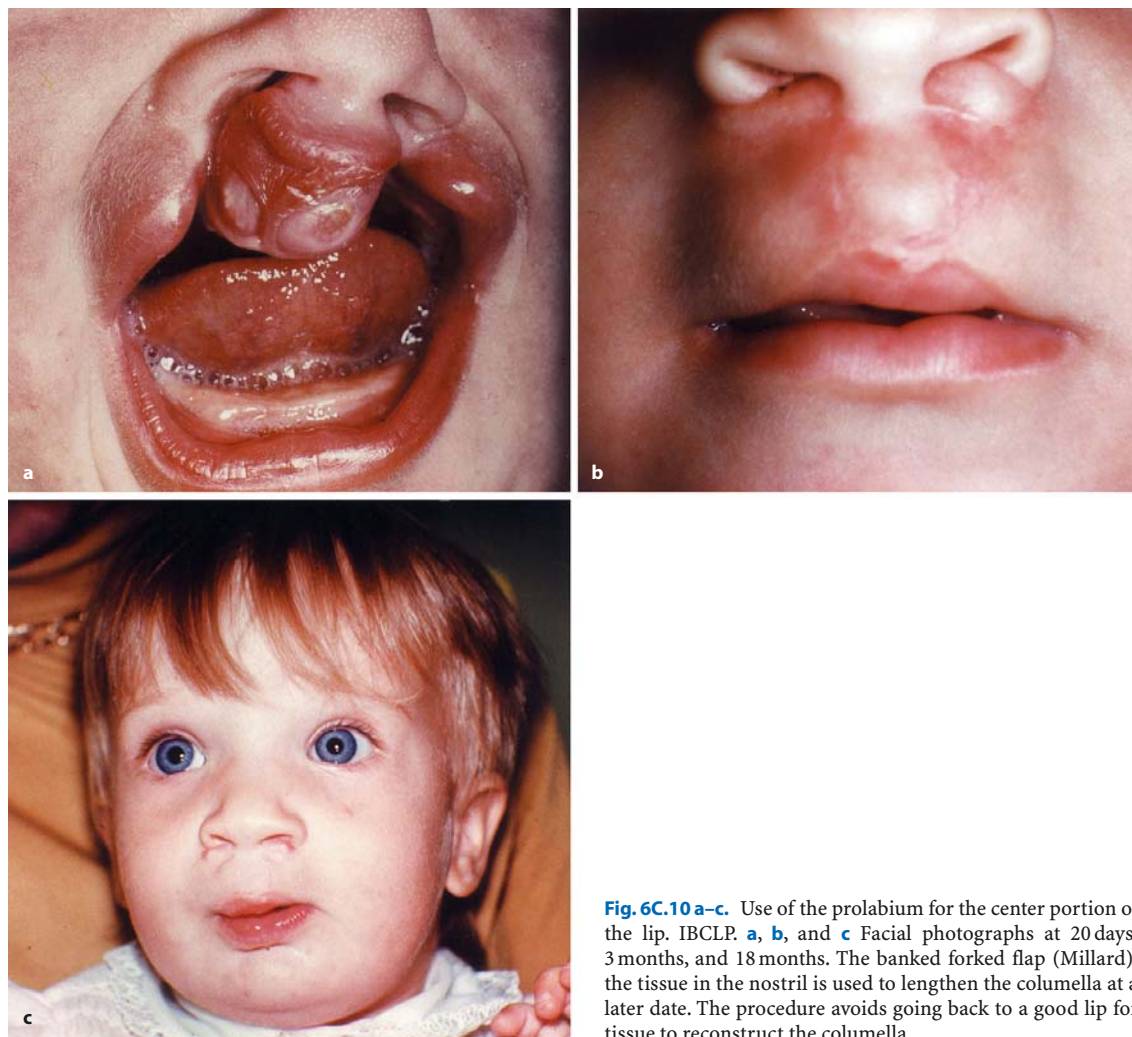


Fig. 6C.10 a-c. Use of the prolabium for the center portion of the lip. IBCLP. **a**, **b**, and **c** Facial photographs at 20 days, 3 months, and 18 months. The banked forked flap (Millard): the tissue in the nostril is used to lengthen the columella at a later date. The procedure avoids going back to a good lip for tissue to reconstruct the columella

Serial profile analyses of cast and cephalographs reported in Berkowitz's earlier studies and the serial studies presented in this text demonstrate that, during the first 2 years following surgery, the united lip exerts a posteriorly directed pressure force on the protruding premaxilla. He speculates that this force is exerted through the nasal septum to the premaxillary vomerine suture, gradually retarding the forward and vertical growth of the nasal septum with the attached premaxilla. The growth of the lateral palatal segments does not appear to be affected by this force. Inhibition of midfacial growth coupled with forward and vertical growth of the upper face and mandible is responsible for the eventual flattening of the facial profile (Figs. 6C.5, 6C.6).

Protrusion of the midface is only slightly reduced by the reduction-remodeling of the labile surface of

the premaxilla associated with the eruption of the permanent incisors. Facial growth data suggest that the tendency toward mandibular prognathism can be considered advantageous, insofar as it reduces the earlier protrusion of the premaxilla. Comparison of the profile changes in children whose lips were united at 6 years with infants whose lips were united soon after birth showed less premaxillary protrusion for children whose lips were repaired in infancy. This suggests that the beneficial effect of lip repair is long-acting in that it continues to exert a restraint on the growth of the premaxillary-vomerine complex long after the lip is repaired [4, 7, 19].

After the lip is united, there are no documented long-term benefits to using intraoral neonatal maxillary orthopedics to control the premaxilla's position relative to the lateral palatal segments (i.e., to inhibit

Fig. 6C.11. Before and after line drawings demonstrating the use of orthopedics for the correction of a malocclusion in a BCLP. 5-9 Frontal and lateral tracings of a child with a bilateral cleft lip and palate before orthopedic treatment. Intracuspisid width was 17 mm; intramolar width was 27 mm; and the interincisal angle was 183°. 7-6 After pre-maxillary advancement and buccal expansion, the intracuspisid width changed to 25 mm, intramolar width to 37 mm, and the interincisal angle to 145°. All changes in measurements were dependent on the bodily (orthopedic) movement of the bony segments and not on the movement of teeth (orthodontics). After the palatal segments are properly aligned, the movement of teeth will follow. While waiting for the permanent teeth to erupt, the new arch form needs to be retained with a fixed appliance

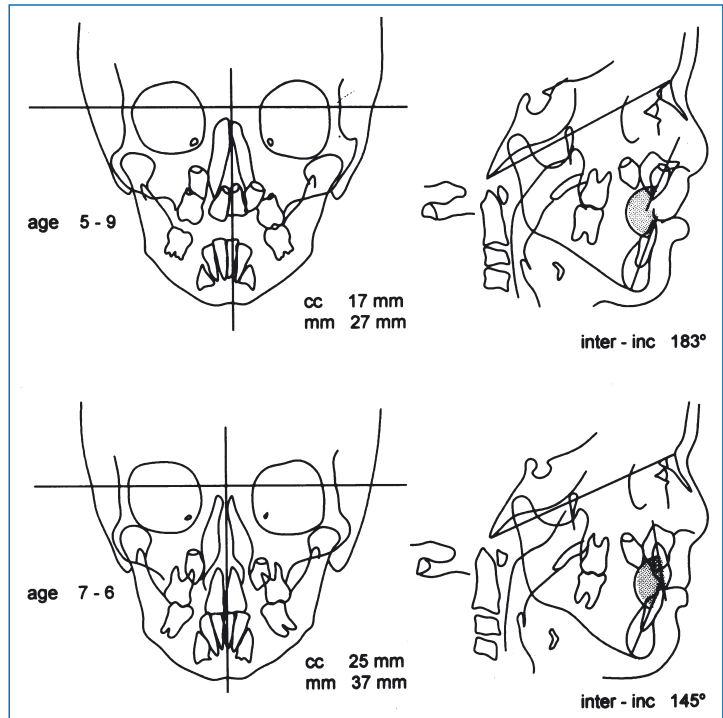


Table 6C.1. Timing and sequencing of surgical-orthodontic treatment (conservative).

Age	Orthodontics	Surgery
After birth	CBCLP-external elastics (off head bonnet) over protruding premaxilla – no obturator	
3–4 weeks		Lip adhesion
6 months		Millard Forked Flap
18–30 months		von Langenbeck ^a (simultaneous closure of the hard and soft palate)
4–5 years	Correction of buccal crossbite only using a fixed quad helix palatal expander	
5–7 years	Fixed palatal retention	
7–8 years	Align anterior teeth prior to secondary alveolar bone graft (SABG)	Secondary bone graft using cranial or iliac crest bone
9–13 years	Full banded treatment with or without maxillary protractor (Delaire faciai mask)	Nasal tip revision
13–17 years	Full orthodontics. Evaluate need for surgical orthodontics (Distraction osteogenesis or Lefort I)	Maxillo-mandibular surgery ^b
17–18 years	Postsurgical orthodontics followed by prosthetics	Nasal-lip revisions

^a Timing of palate closure depends on the width of the cleft space relative to the adjoining palatal surface area. Anterior cleft is left open.

^b Maxillo-mandibular surgery is usually performed earlier for females (around 15–16 years of age) than males whose surgery is performed during the summer prior to the senior high school year. This allows for maximum facial growth changes and leaves enough time to perform postsurgical orthodontics and prosthetic treatment.



Fig. 6C.12 a–f. Millard's surgical lip procedure uses the probulum to construct the entire midportion of the lip over the protruding premaxilla. This case was selected to show an example to severe upper lip protrusion that can result due to a

severely protruding premaxilla. This case and other presented in this chapter will show how the face improves with time, that is, facial growth

the palatal segments from spontaneously moving together), nor is it important that the premaxilla be accommodated within the arch at this time. There is no evidence that overlapping palatal segments suffer growth inhibition. In most cases, proper premaxillary alignment within the lateral palatal segments can be easily achieved with orthodontics in the deciduous or mixed dentition without resorting to neonatal maxillary orthopedics. The following cases show that orthodontics in the permanent dentition, in the absence of growth-inhibiting palatal surgery, and with a good or poor facial growth pattern, eventually will lead to ex-

cellent facial aesthetics and dental function. Any resulting buccal crossbite can be easily corrected at 4 or 5 years of age, when the child is manageable in a dental chair, using fixed tooth-borne or even removable arch expansion appliances. The author strongly favors the use of fixed appliances for arch expansion and retention.

There are many occasions when the profile shows midfacial retrusion, due to severe ventroflexion of the premaxilla. As long as during the mixed dentition gingivoperiosteoplasty has not been performed (see Chap. 20), the premaxilla can easily be brought for-

ward orthodontically and held in place until alveolar bone grafting is performed. Retention of the corrected maxillary arch is still necessary until orthodontics are completed (Fig. 6C.11).

6C.6.1 Why Some Premaxillae Continue to Project Following Lip Repair and Others Do Not

The cases being presented will show that there are a number of treatment planning facial factors. For example, the patient's facial growth pattern and the amount of palatal osteogenic deficiency are beyond the control of the surgeon. The integrated growth of the entire face is important to resolve the profile deformity. When the end result is unfavorable – that is, the facial profile remains highly convex – the premaxilla and the body of the maxilla usually have grown forward with limited forward mandibular projection or tension has been created at the PVS, causing further premaxillary protrusion.

Facial growth studies by Handelman and Pruzansky [12] and by Berkowitz (presented in this chapter) have shown that, in highly convex facial profiles with a moderately protrusive premaxilla and a recessive mandible, the lip musculature usually is hypertonic and positioned over the upper incisor teeth, resulting in severe premaxillary ventroflexion. In mesognathic faces with isotonic or hypotonic lip musculature and a severely projecting premaxilla, the lower lip may be positioned lingual to the premaxilla and between the upper and lower incisors, creating an anterior component of force at the premaxillary vomerine suture (PVS). This may cause additional bony growth at the suture, leading to greater premaxillary protrusion with a normal maxillary incisal axial inclination. The lower incisors may be tipped lingually (Figs. 6C.11, 6C.12).

The serial record present in the case reports to follow will show that it can be anticipated that in most cases (53 out of 59 CBCLP; S Berkowitz, unpublished data) good faces will have developed at adolescence. Figure 6C.14 shows serial computer-generated CBCLP palatal cast outlines which were superimposed on the palatal rugae and registered on the vomer. All of these cases had the palate closed between 18–24 months using a von Langenbeck with modified vomer flap (it was readily observed at every age period).

This illustration clearly demonstrates that the premaxilla is in the same place in the palate at adolescence as it was at birth. It is retained in place by the surrounding facial musculature in most cases. There are exceptions, usually dependent on the balance of the facial musculature.

6C.6.2 Dental Occlusion

Analyses of the dental occlusion of many patients with complete bilateral clefts of the lip and palate at 6 years of age rarely show retruded maxillary shelves (i.e., a Class III relationship) [7]. The buccal teeth are most often in a Class I or 2 relationship. A Class III relationship exists only if the maxilla is retrusive due to severely retarded growth caused by traumatic surgery and/or the patient's phenotype. The mandible is rarely prognathic at this age. In many cases, due to palatally osteogenic deficiency and severe dental crowding in the maxillary arch, some teeth may need to be extracted. This may or may not have to be done in the mandibular arch. There is conclusive evidence to suggest that the cleft palatal shelves may be deficient in mass, but there is no evidence that they have lost their growth impetus from having been detached from the growing nasal septum.

Until proven otherwise, the surgeon should have confidence that the severe facial convexity seen at birth can diminish with time as the united lip restrains the forward development of the midface while the mandible increases in size and is positioned more downward and forward relative to the growing anterior cranial base and midface.

6C.6.2.1 After Birth

In general, no presurgical orthopedics were provided other than the use of a head bonnet. In complete bilateral cases only, an external facial elastic may be utilized to ventroflex the protruding premaxilla to reduce tension at the surgical site when utilizing lip adhesion. External traction is usually not necessary in incomplete clefts of the lip and palate.

The Forked Flap (Fig. 6C.13)

All of the bilateral cases were treated with Millard's Forked Flap [54]. It was designed to lengthen the columella as a primary or secondary procedure. Advantages include:

- Release of depressed nasal tip
- Lengthen the short columella
- Reduce an unattractive wide prolabium
- Revision of bilateral lip scars
- Reduction of flaring alar bases

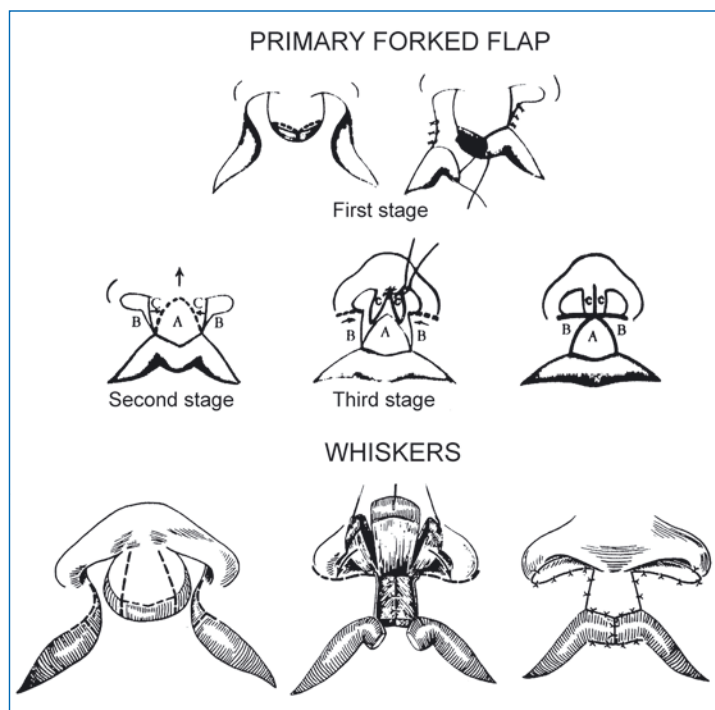


Fig. 6C.13. In 1956–1957 Millard designed a secondary forked flap for columella lengthening. He believed at times it is best to take a little tissue from the prolabium and the lip and store it in the area for later use in reconstructing the columella. The forked flap was originally designed as a secondary procedure; however, it can be used as a primary procedure when the columella is extremely short and the prolabium is of reasonable size. *Top:* In the primary forked flap, the fork flap is elevated and advanced into the columella with release of the nasal tip. *Bottom:* The whisker fork flap involves joining the lip muscle and banking the fork. The alar bases are joined together in the midline and the forks partially tubed on themselves and led into the transverse incisions between the lip and alar bases, whisker fashion. This surgery is delayed several years. Millard likes this procedure best of all

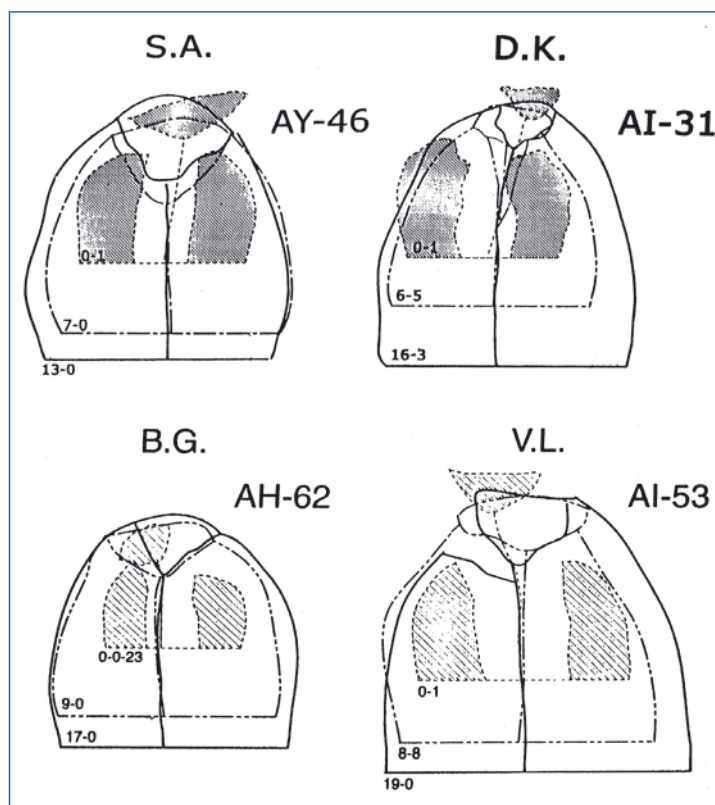


Fig. 6C.14. Computerized digital tracings of the perimeter of the outline of CBCLP palatal casts superimposed on the palatal rugae and registered on the vomer. Palatal cleft closure between 18 and 24 months using a von Langenbeck and vomer flap. Note that the premaxilla is held in place while the palate grows in all directions; posterior palatal growth occurs to accommodate the developing molars. None of these cases were treated with presurgical orthopedics

AXIAL INCLINATION OF MAXILLARY CENTRAL INCISOR (Imx to SN)

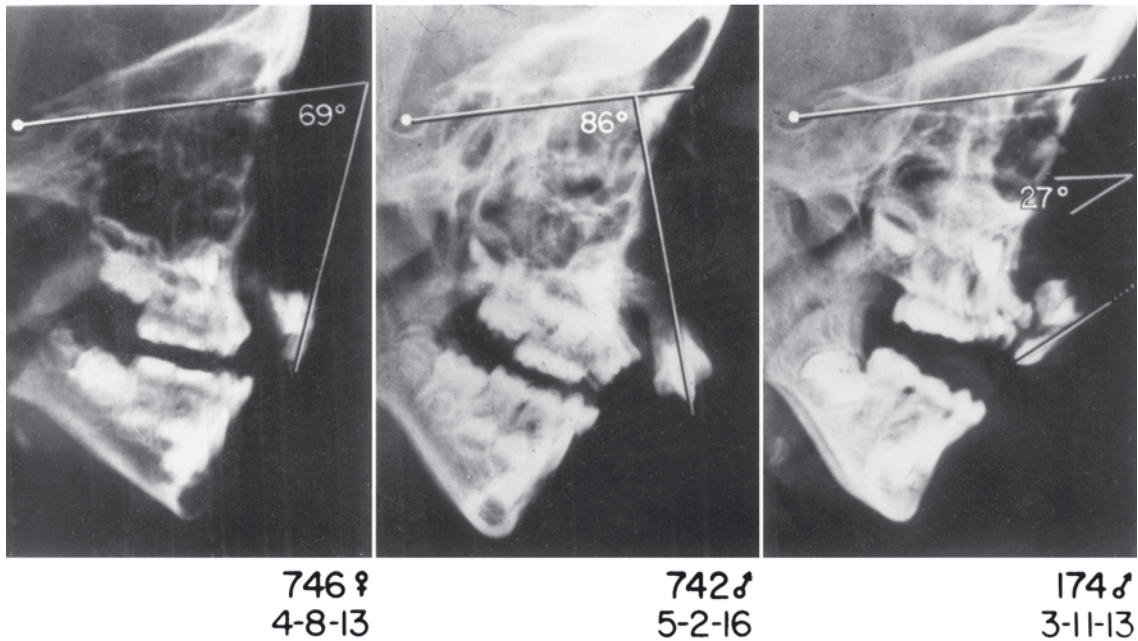


Fig. 6C.15. In spite of the lip surgery performed, the position of the premaxilla in the anterior dentition is determined by the facial pattern and tonicity of the surrounding facial musculature. *Case #746:* Protruding premaxilla surrounded by hypotonic lip musculature. *Case #742:* Retrognathic mandible with protrusive premaxilla. The lower lip is positioned between the upper and lower incisors creating a forward force at the PVS.

The resulting increase in tension at the PVS stimulates additional growth and causes increase in inclination of the upper incisors. *Case #174:* Retrognathic mandible with less premaxillary protrusion hypertonic lip musculature creating severe lip pressure which ventroflexes the premaxilla. Variations in premaxillary position is unpredictable [12]

Millard [54] suggests that the primary fork flap procedure is not appropriate for all bilateral cleft cases. Whether to use the procedure depends on: (1) The position of the premaxilla. It should not be used when the premaxilla is severely protruded. (2) The size of the prolabium. The width of the prolabium determines whether the flap is possible, and the vertical length indicates the amount of columella lengthening available. (3) Columella length. This discrepancy must be measured not only in actual length in millimeters

of the columella but an estimation of the patient's desired final length must also be made.

There are occasions where the forked flaps from the prolabium are banked with a subalar incision, whisker fashion. The alar base is not advanced medially in an attempt to leave a subalar gap in which to store the forks. Millard delays shifting of the banked forked flap into the columella for several years (from 6 months to 6 years).



Fig. 6C.16. Early premaxillary surgical setback and palatal cleft closure at 7 months of age. The lateral lip elements were brought together below the prolabium. Results at 7 years of age: (1) long tight upper lip, (2) anterior open bite due to failure of the premaxilla to descend with the palate, and (3) the surgery (push back) used to close the palatal cleft at 7 months obliterated the vault space interfering with tongue posture. The tongue is, therefore, being carried forward with the tongue tip protruding, preventing the incisors from reaching the occlusal plane



Fig. 6C.17 a-c. The lateral lip elements were brought below the prolabium creating a long tight lip

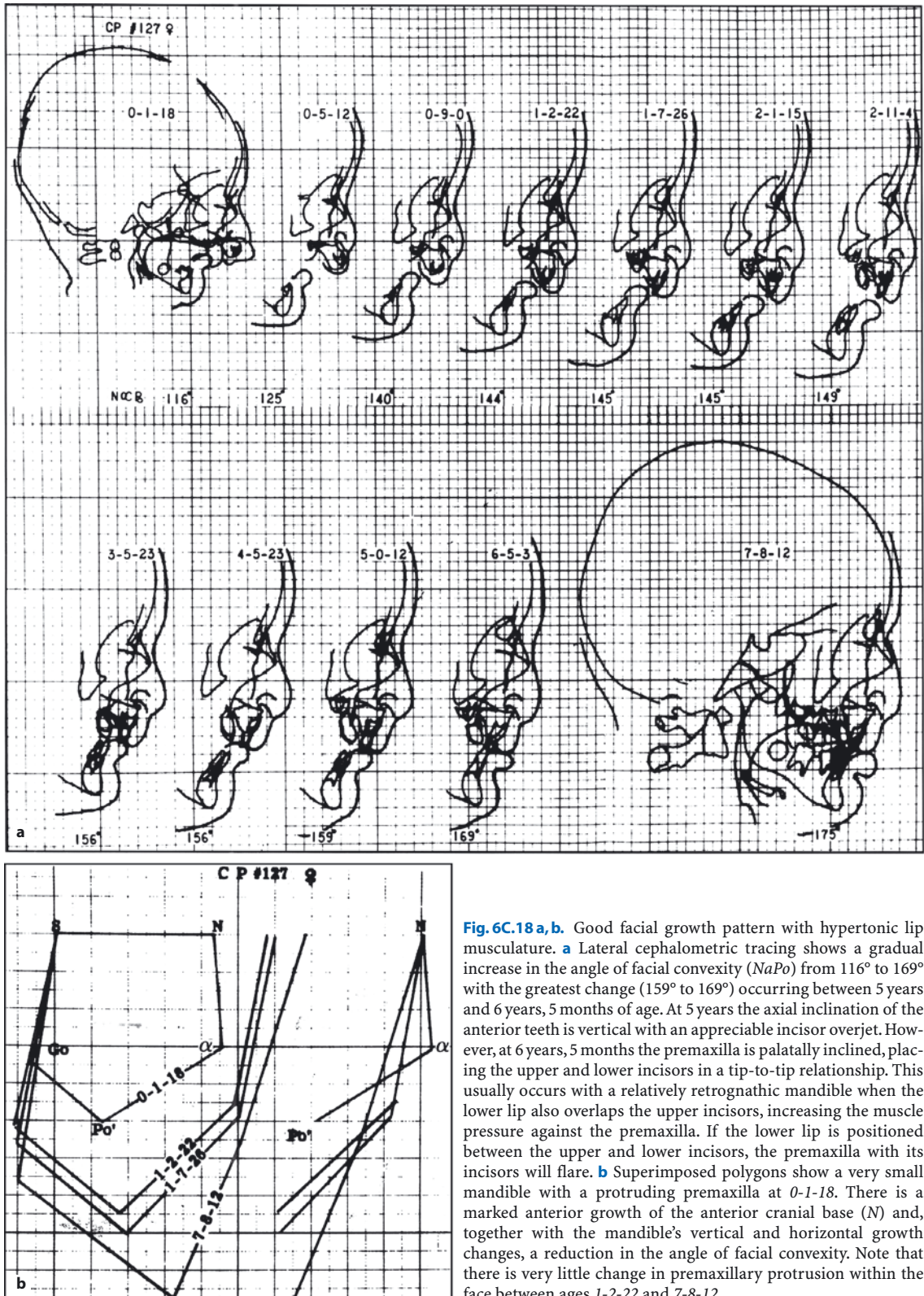


Fig. 6C.18 a,b. Good facial growth pattern with hypertonic lip musculature. **a** Lateral cephalometric tracing shows a gradual increase in the angle of facial convexity ($NaPo$) from 116° to 169° with the greatest change (159° to 169°) occurring between 5 years and 6 years, 5 months of age. At 5 years the axial inclination of the anterior teeth is vertical with an appreciable incisor overjet. However, at 6 years, 5 months the premaxilla is palatally inclined, placing the upper and lower incisors in a tip-to-tip relationship. This usually occurs with a relatively retrognathic mandible when the lower lip also overlaps the upper incisors, increasing the muscle pressure against the premaxilla. If the lower lip is positioned between the upper and lower incisors, the premaxilla with its incisors will flare. **b** Superimposed polygons show a very small mandible with a protruding premaxilla at 0-1-18. There is a marked anterior growth of the anterior cranial base (N) and, together with the mandible's vertical and horizontal growth changes, a reduction in the angle of facial convexity. Note that there is very little change in premaxillary protrusion within the face between ages 1-2-22 and 7-8-12

6C.6.2.2 In the Deciduous Dentition (3–6 Years of Age)

The purpose of treatment at this age is to align the palatal segments in order to obtain a more normal contour to the alveolar ridge, reshape the palatal vault, and provide a more symmetrical foundation for the support of the lips and nose. These changes are possible because orthodontic treatment in cleft palate allows for the movement of palatal segments when using a von Langenbeck procedure in addition to altering the position of the teeth within the alveolus. Because the roof of the mouth is also the floor of the nose, realignment of the palatal processes not only produces desirable alteration in the contour of the palate, but also induces similar changes within the floor of the nose.

Most clinicians agree that nontraumatic conservative surgery will not solve all problems for all complete bilateral cleft lip and palate cases. Although buccal crossbites can be corrected in the deciduous dentition, some advocate orthodontic/orthopedic repositioning of the premaxilla at a later age (in the mixed dentition prior to or after secondary alveolar bone grafting) when the facial growth pattern leaves no alternative and dictates that it is the procedure of choice. Some orthodontists prefer to do this in the permanent dentition.

6C.6.2.3 Mixed Dentition (6–11 Years of Age)

When a child with a cleft starts elementary school, even if the protruding premaxilla still extends far forward of the lateral palatal segment creating a severe convex facial profile, a surgical premaxillary setback should be considered solely for aesthetic reasons because there can be long-term deleterious effects. This decision is a difficult one. One is damned if one does or doesn't! One has to balance the child's social and psychological needs with long-term facial developmental factors.

In the last 20 years, many reports published with improved documentation were critical of surgical premaxillary setback performed in the newborn period, or even in early adolescence prior to the prepubertal growth spurt. Fortunately, these criticisms led to abandonment of this procedure [19].

Orthodontics applied during the mixed dentition, in preparation for or after secondary alveolar bone grafting (the placement of bone in the alveolar cleft after the first year of age), can aid in reducing the premaxillary overbite and maintaining the premaxilla within the lateral palatal segments. Eliminating the alveolar cleft permits the unerupted teeth (lateral in-

cisors and cuspids) adjacent to the cleft to erupt or be moved into proper position.

In Berkowitz's experience, a protruding premaxilla – with or without a large anterior cleft space – at 6–7 years of age does not signify that the same unpleasant aesthetic conditions will persist into adolescence. After the pubertal facial growth spurt, when the facial convexity is markedly reduced because of increased mandibular and upper facial growth, facial aesthetics will be greatly improved in most patients.

There is strong psychosocial pressure on clinicians to improve facial aesthetics as soon as possible, even when the premaxilla cannot be repositioned in the absence of an anterior cleft space. For this reason, many unsuccessful attempts have been made to surgically or mechanically set the premaxilla back at an early age in the hope of preventing an unaesthetic midface at a later age, believing the midfacial growth would still be normal. At this difficult period, every effort should be made to convince both the parents and the child, using before and after photographs, that facial aesthetics will greatly improve as the face grows and that the long-term benefits far outweigh any early improvements at the expense of future good facial growth.

6C.6.2.4 At Adolescence

If a poor facial growth pattern at adolescence with a large anterior cleft space leads to a protruding premaxilla set well forward in the facial profile and the mandibular incisors, the premaxilla can be surgically set back. A secondary alveolar bone graft is simultaneously performed to close the remaining anterior cleft space. A fixed palatal retainer or a heavy labile arch wire must be utilized for at least 2 months as a supporting splint for the three segments during the healing period.

There are cases in which both buccal segments are in a Class I relationship, a large anterior cleft space is present, and the premaxilla is missing one or both lateral incisors, yet it is in an ideal overjet and overbite relationship. If the surgeon believes that the anterior cleft space is too large to be successfully closed by a secondary alveolar bone graft, the only remaining option is to surgically advance one or both palatal segments, placing the buccal teeth in a Class II relationship with at least one, but sometimes both, of the cuspids in the lateral incisor space. This usually necessitates the surgical increase in posterior transpalatal width. A secondary alveolar bone graft is performed at the same time [55].

There are too many variations of orthodontic problems to review each of the treatment plans separately. However, in most cases, a common treatment sequence is usually followed. First, the posterior palatal crossbite needs to be orthopedically corrected in the deciduous or mixed dentition using quadhelix mechanics. Reducing maxillary arch crowding by tooth extractions may be necessary due to the lack of palatal osteogenic tissue. Achieving ideal anterior teeth aesthetics often requires that the lost lateral incisor spaces be recovered for the eruption of impacted lateral incisors or cuspids. In most cases, establishing room for four maxillary incisors by expansion and slight incisor advancement is the ideal treatment. A slightly retrusive maxilla with mild crowding may require the advancement of the upper jaw using protraction forces for at least 6–12 months to obtain an ideal anterior overbite-overjet relationship. In some instances, the extraction of one lower incisor to correct the mandibular anterior arch will be necessary to reduce an anterior crossbite.

In all cleft types, involving the lip and palate, orthodontic surgical treatment decisions are often dictated by the relationship of the premaxilla to the lateral palatal segments and the facial growth pattern (i.e., whether the face is prognathic, mesognathic, or retrognathic). For example, if a slightly retruded maxilla and a retrognathic mandible coexist, it may be necessary to surgically advance the lower jaw with or without doing the same to the maxilla. In most cases, orthopedic protraction to move the maxilla should be considered first.

If one or both maxillary buccal teeth are in a Class II relationship and one or both lateral incisors are absent, it is advantageous to move one or both cuspids into the lateral incisor space after secondary alveolar bone grafting. Eruption of the maxillary cuspids with its supporting alveolar bone usually closes off any remaining anterior cleft space. In very rare instances, when a large anterior cleft space exists and there is insufficient mucoperiosteum to close this space and a good anterior overbite-overjet relationship exists, the treatment of choice is to advance one or both palatal segments to make contact with the well-positioned premaxilla. A secondary alveolar bone graft can be performed at the same time [55] (Fig. 6C.59).

After the pubertal facial growth spurt, the once protruding premaxilla usually is no longer an aesthetic problem. Not surprisingly, at this age the midface may be retruded even if the premaxilla was not surgically or mechanically set back and especially if there is good upper and lower facial growth. Treatment now needs to be focused on advancing the midface and/or the premaxilla using either orthopedic forces from a protraction facial mask or a maxillary osteotomy (LeFort I advancement).

In 30 years of clinical experience, surgical premaxillary retropositioning was performed only twice and only for aesthetic demands. It was never necessary to reduce its protrusion at the adult stage. Facial growth (time) ultimately is the surgeon's and orthodontist's best friend – and a most trenchant critic.

6C.6.2.5 Retention

Permanent retention of the maxillary arch form in all clefts of the lip, alveolus, and palate is sometimes necessary even after secondary alveolar bone grafting, depending on the extent of transpalatal scarring and lip-cheek pressure. It requires either a removable palatal prosthesis or preferably a fixed bridge spanning the cleft. Arch collapse with a return of buccal and/or anterior crossbite can occur rapidly in the presence of extensive lip and transpalatal scar tissue, because scar tissue has limited stretch capability.

6C.7 The Following Case Studies Represent Conservative Surgical and Orthodontic Treatment Sequence

Cleft lip and cleft palate defects offer a degree of habilitation that is not available for many equally serious congenital or acquired handicaps. Achieving this potential requires careful planning and skillful execution of treatment that respects individual growth patterns (Table 6C.1). The following cases were selected to show various treatment procedures, some of which were unsuccessful but nevertheless have teaching value because they reflect on physiological principles (Figs. 6C.19–6C.73, Tables 6C.2–6C.4).



Fig. 6C.19 a-i. Case TM (WW-9) demonstrates excellent facial and palatal growth changes in an incomplete bilateral cleft lip and palate treated conservatively (no PSOT). **a** Newborn. **b** At 7 months, after lip closure. **c** 4 years, 2 months. Note buccal crossbite on the left side. The crossbite was corrected by 4 years, 7 months using a fixed palatal expander. **d** At 8 years after the central incisors were aligned and a secondary alveolar bone graft was performed at 7 11/42 years. Note that lateral incisors are erupting through the graft. **e** Palatal view at 7 11/42 years. **f** Full face at 17 years. **g** Close-up of lip. **h** Occlusion at 17 years after orthodontics. **i** Palatal view showing good vault space with minimal scarring

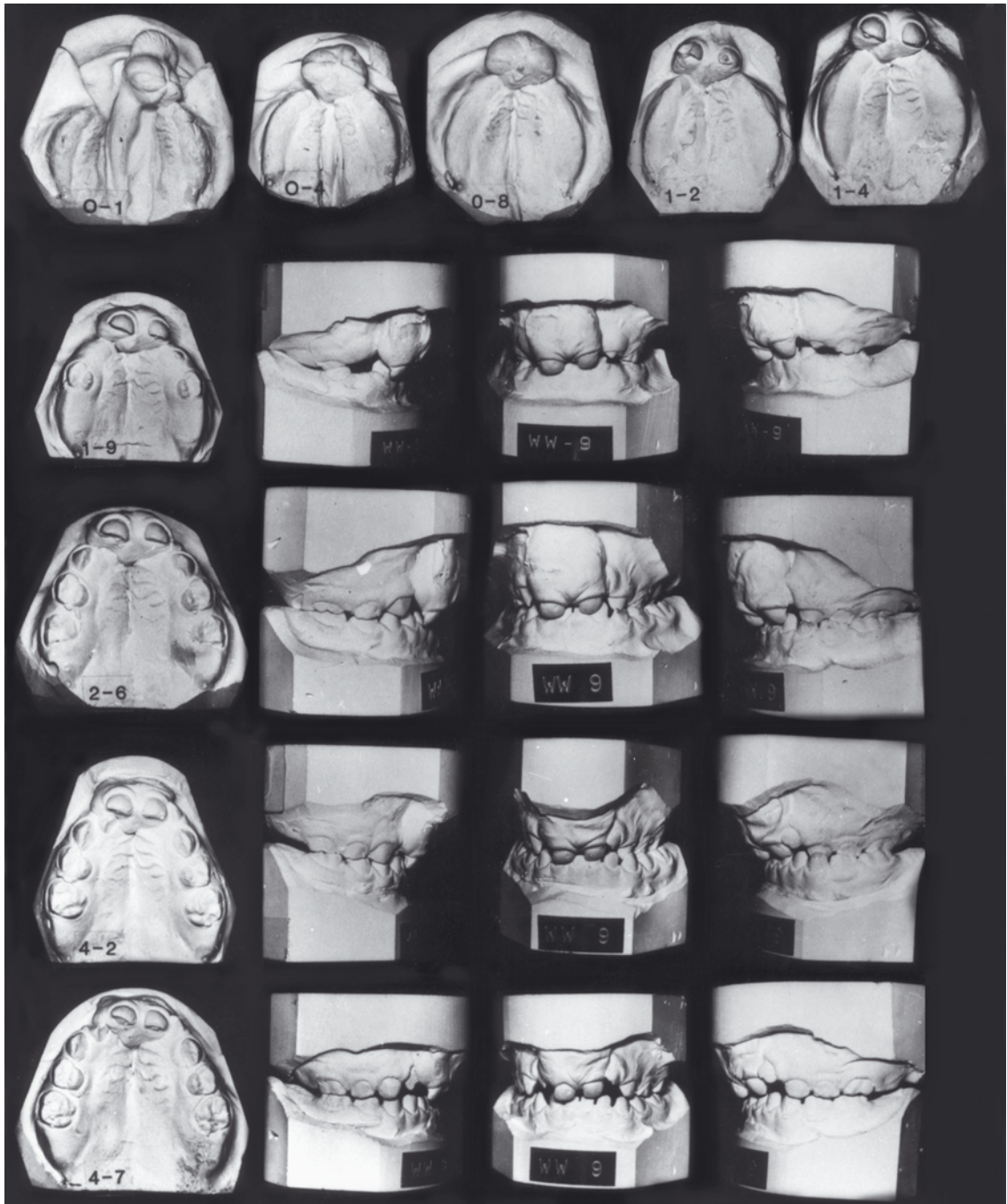


Fig. 6C.20. Case TM (WW-9). Serial palatal cast changes when no presurgical orthopedics were utilized. 0-1 Newborn, prior to lip surgery. 0-4 After lip adhesion the lateral palatal segment moved medially behind the premaxilla. 0-8 The cleft space is much smaller and continues to decrease in size with palatal

growth. The premaxillary position at this age poses no treatment or growth problem. 1-2 The palatal cleft was closed with a modified von Langenbeck procedure. 1-9 Ideal incisal overbite and overjet. 2-6 and 4-2 Left buccal crossbite which was corrected at 4-6 using a fixed palatal expander. 4-7 Good occlusion

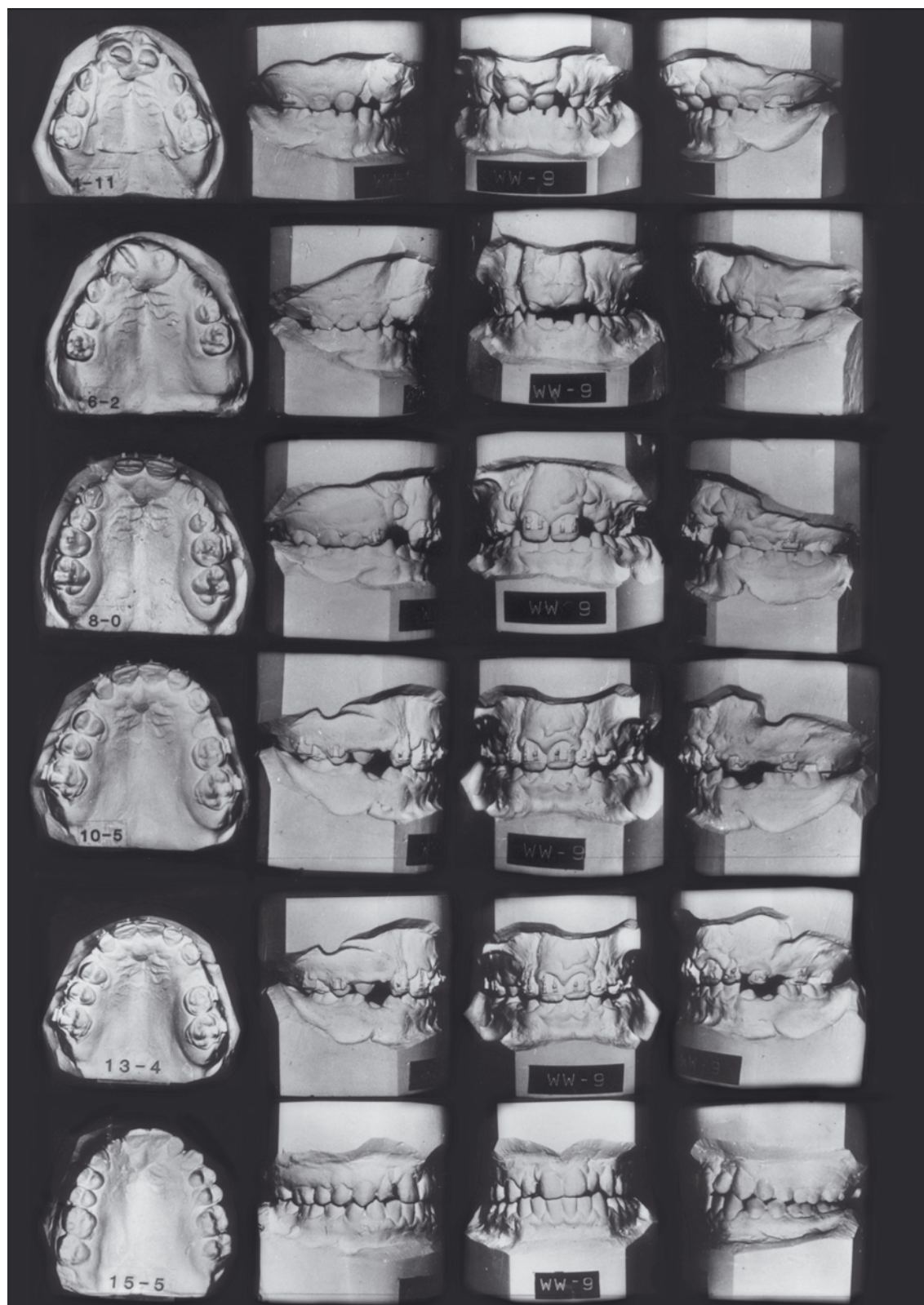


Fig. 6C.20. (continued)

Fig. 6C.20. (continued) 4-11 Outline of fixed palatal retainer is seen on the case. Note that the premaxilla is now positioned within the palatal segments. 6-2 The central incisors in the area of the cleft are frequently found to be rotated. 8-0 Six months after a secondary alveolar cranial bone graft. The right and left

lateral incisors have erupted through the graft. 10-5 The lateral incisors are aligned with the central incisors. 15-5 Excellent occlusion after orthodontia. A removable maxillary retainer is being used to maintain the arch form

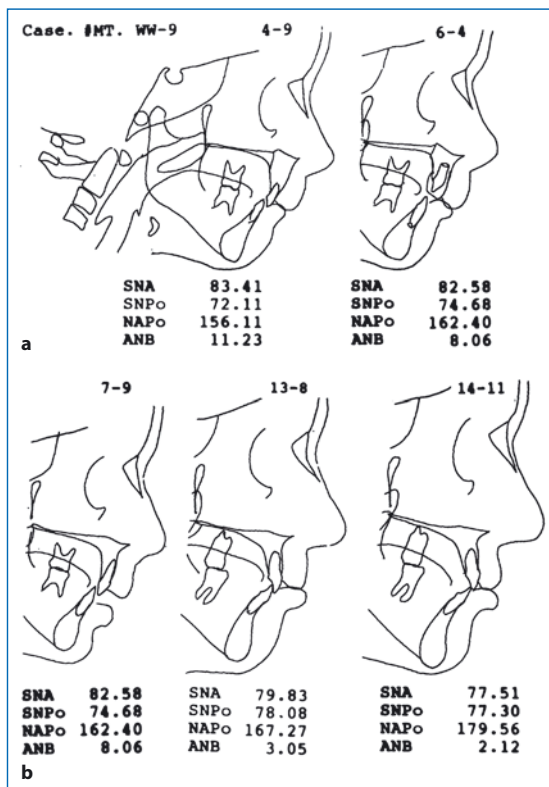


Fig. 6C.21 a,b. Case TM (WW-9). This series depicts an excellent facial growth pattern. **a** Computerized tracings of facial skeletal and soft tissue profile changes. At 14-11, the profile measurements are well within the normal range. **b** Facial polygons superimposed on the SN line and registered at S demonstrate that the excellent facial growth pattern reflects some anterior cranial base growth, very little forward growth of the midface, and excellent mandibular growth in a downward and forward direction. Note that the midface was still protrusive at 7-9, but after the mandibular pubertal growth spurt (13-8), the facial profile flattened markedly

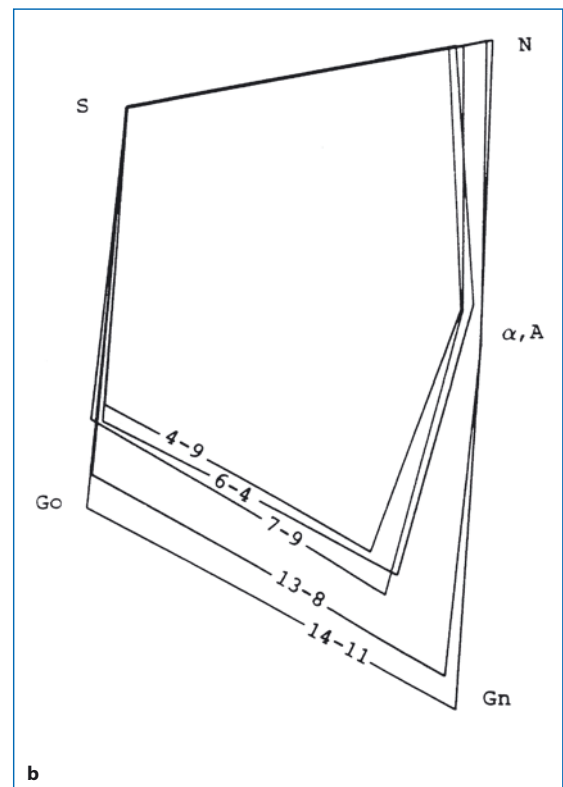


Fig. 6C.22. Case TM (WW-9) Computerized tracings of facial skeletal and soft tissue profile changes. At 14-11, the profile measurements are well within the normal range. **b** Facial polygons superimposed on the SN line and registered at S demonstrate that the excellent facial growth pattern reflects some anterior cranial base growth, very little forward growth of the midface, and excellent mandibular growth in a downward and forward direction. Note that the midface was still protrusive at 7-9, but after the mandibular pubertal growth spurt (13-8), the facial profile flattened markedly

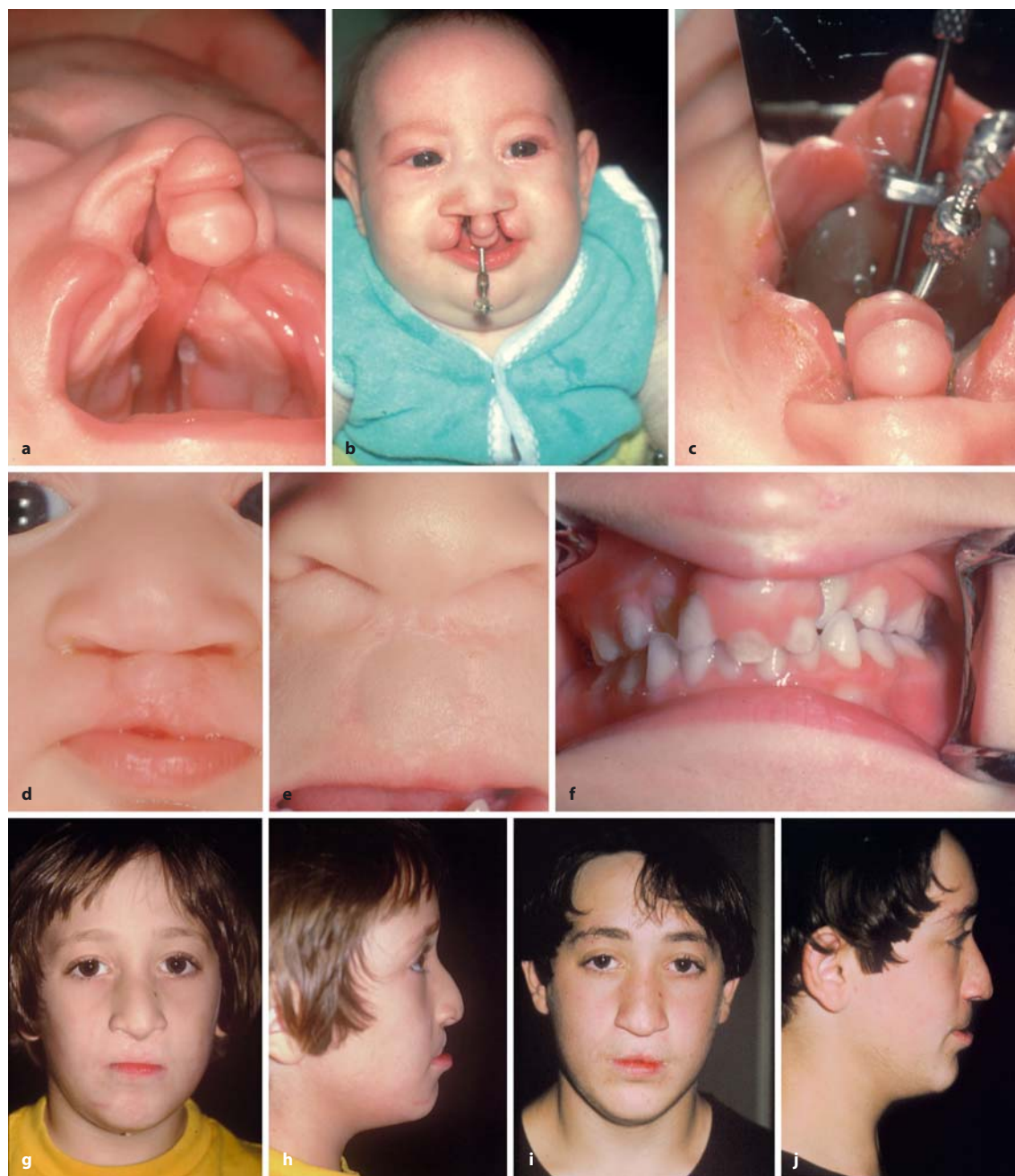


Fig. 6C.23 a–q. Case DK (AI-31) demonstrates unsuccessful use of Latham’s presurgical premaxillary mechanical retraction procedure in a CBCLP, which then required the use of external facial elastics prior to lip surgery. **a** At birth, note the very small protruding premaxilla. **b** Facial photograph with Millard-Latham (M-L) mechanical premaxillary retraction appliance in place. **c** Intraoral view of M-L appliance in place. The appliance was not able to retract the premaxilla due to its small size and was discarded. **d** After wearing a head bonnet with an elastic strap for 2 weeks, the lip was united over the premaxilla. At 2 months the nasal tip is severely depressed. **e** At 3 years of age, the banked tissue of the “forked flap” waiting to be placed in the

columella. **f** 5 years of age. Note excellent occlusion. Radiographs showed that there were no permanent incisors in the premaxilla. **g** and **h** At 8 years of age, the nasal tip has been elevated, but there is poor upper lip support due to the now retruded premaxilla. Lower right and left first bicusps were extracted at 13 years to reduce the anterior crossbite. **i**, **j**, and **k** At 15 years of age, the mandible is growing forward at a more rapid rate and degree than the maxilla. The earlier buccal and anterior crossbites were corrected and two lateral incisors from the lateral palatal segments were brought into position, improving facial aesthetics



Fig. 6C.23 a–q. (continued) **l–q** The maxillary anterior deciduous teeth were extracted and an anterior bridge fabricated to improve dental function and aesthetics

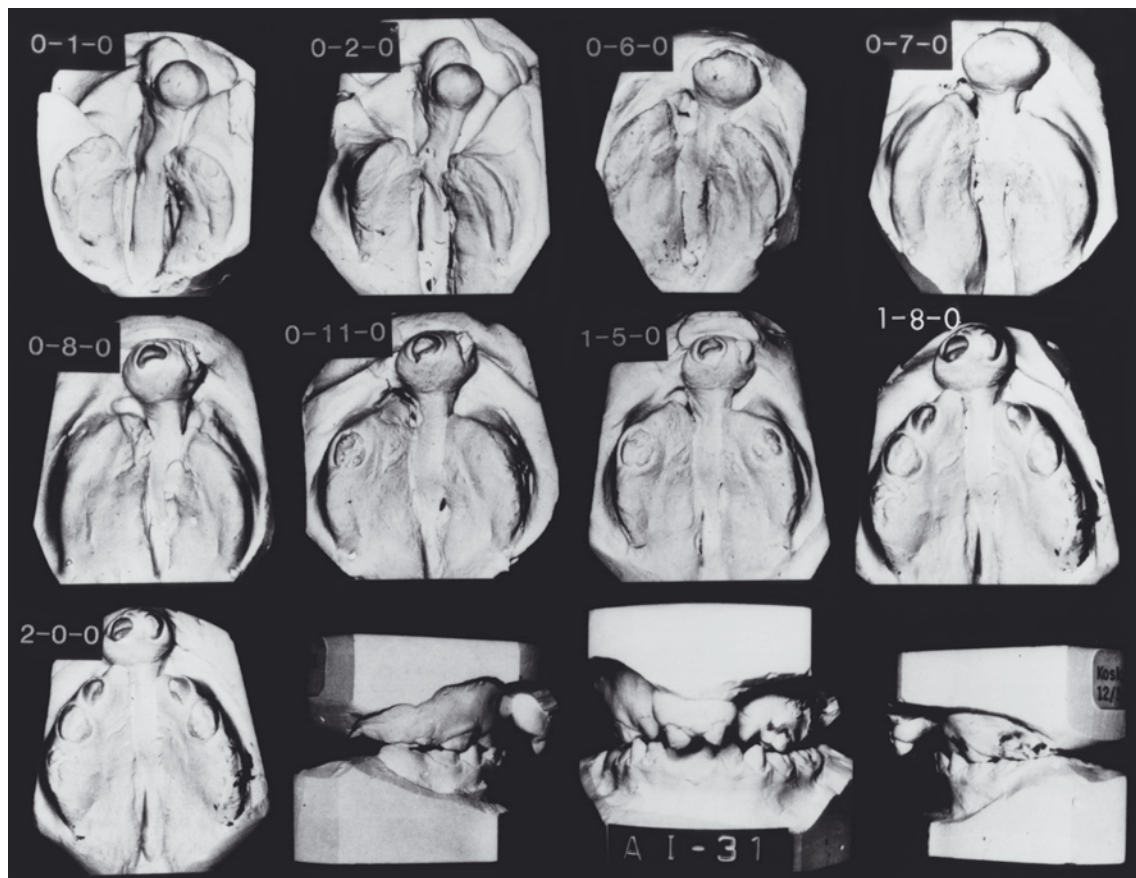


Fig. 6C.24. Case DK (AI-31). **a** Serial dental casts. 1-1-0 and 0-2-0 Note the severely protruding and small premaxilla. 0-2-0 and 0-6-0 The premaxilla ventroflexed under the influence of extraoral elastics attached to a head bonnet. 0-7-0 Marked in-

crease in palatal size coupled with a big reduction in the anterior or cleft space. 2-0-0 Class II right and Class I left occlusion with a severe anterior overjet. Premaxillary surgical setback should not be performed

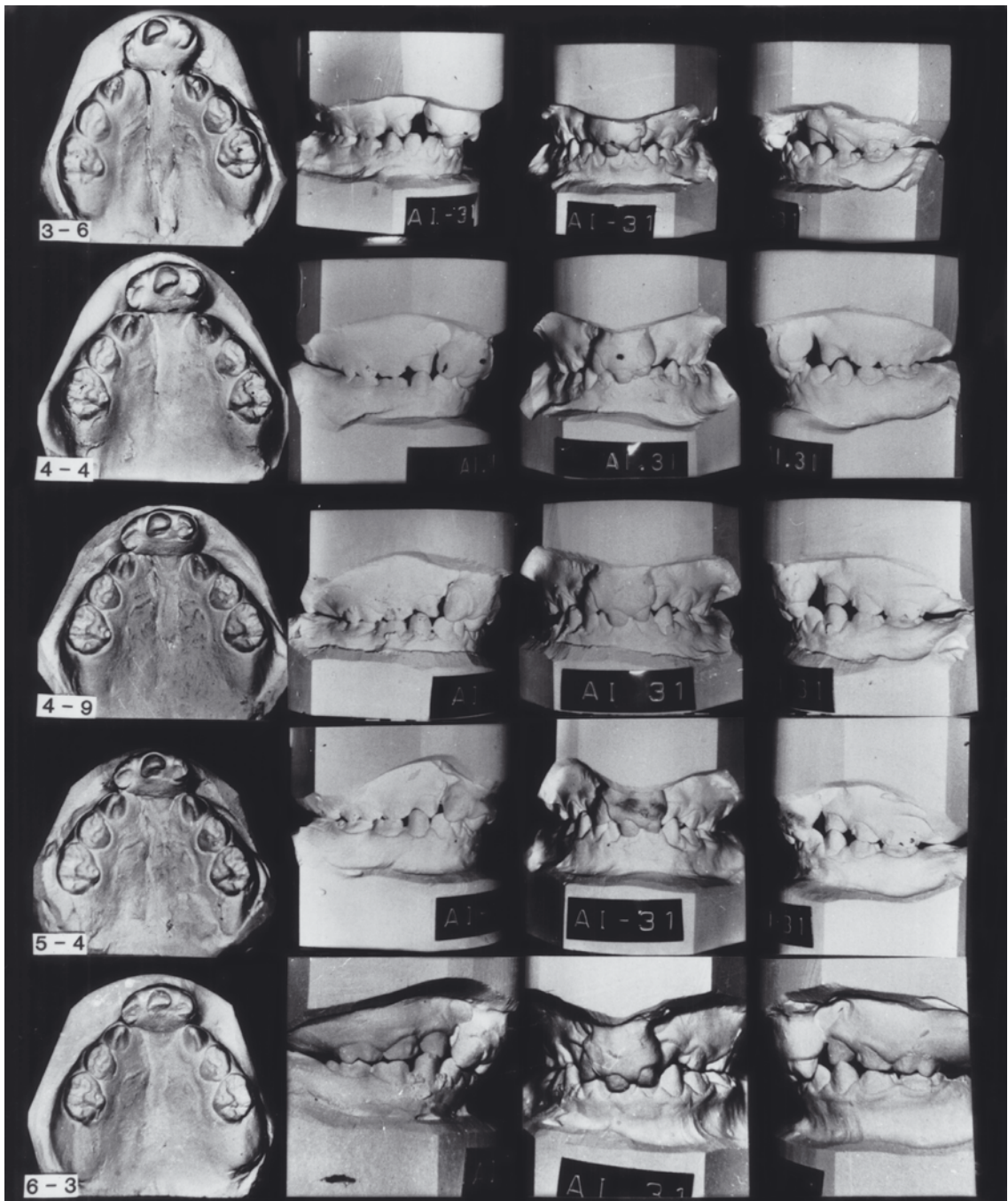


Fig. 6C.24. (continued) 3-6 Note only a small overjet and overbite with a mesioangular rotation of the lateral segments placing the cuspids into crossbite. 4-4 The premaxilla is forward of the lateral palatal segments, with a normal dental overbite/overjet is normal. This occlusal relationship remained for the

next 5 years. The anterior cleft space is left open waiting for additional palatal growth. The anterior palatal cleft did not pose a speech or feeding problem and would allow for palatal expansion

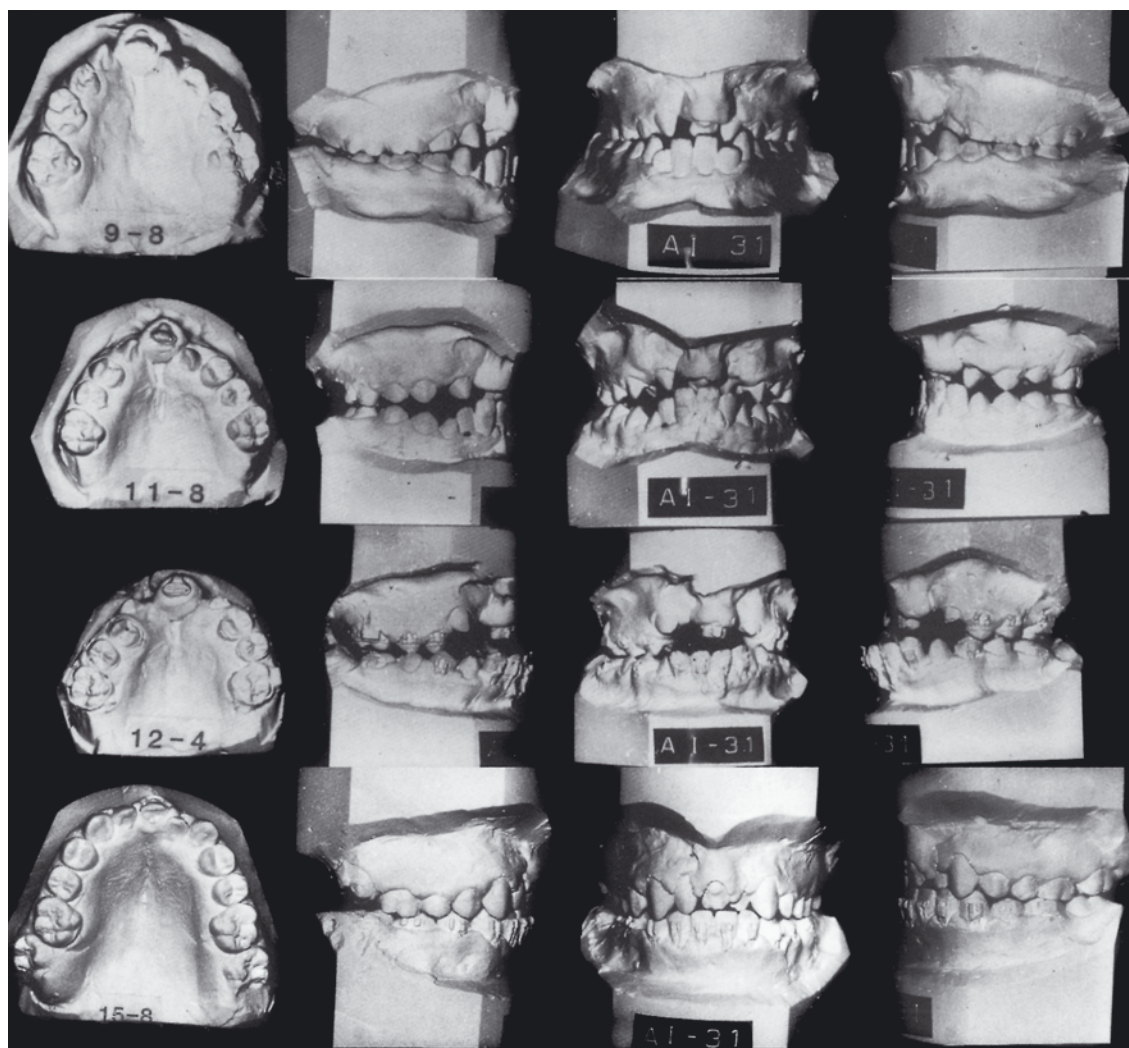


Fig. 6C.24. (continued) 9-8 The remaining deciduous central incisor is now in an anterior crossbite associated with the lack of midfacial development. 12-4 The lower anterior teeth were unsuccessfully advanced to reduce the arch crowding. The excessive flaring necessitated extraction of the lower first bicus-pids. This was followed by retraction of the incisor teeth and space closure with reduction of the severe anterior crossbite.

15-8 Right and left deciduous lateral incisors are brought into position; however, an anterior arch discrepancy still remains. Because of the poor root development of the deciduous lateral incisors, they later were extracted and replaced with an anterior fixed bridge. Comment: This case clearly demonstrates that the anterior occlusion cannot be predicted at birth, therefore, early premaxillary surgical setback should not be performed

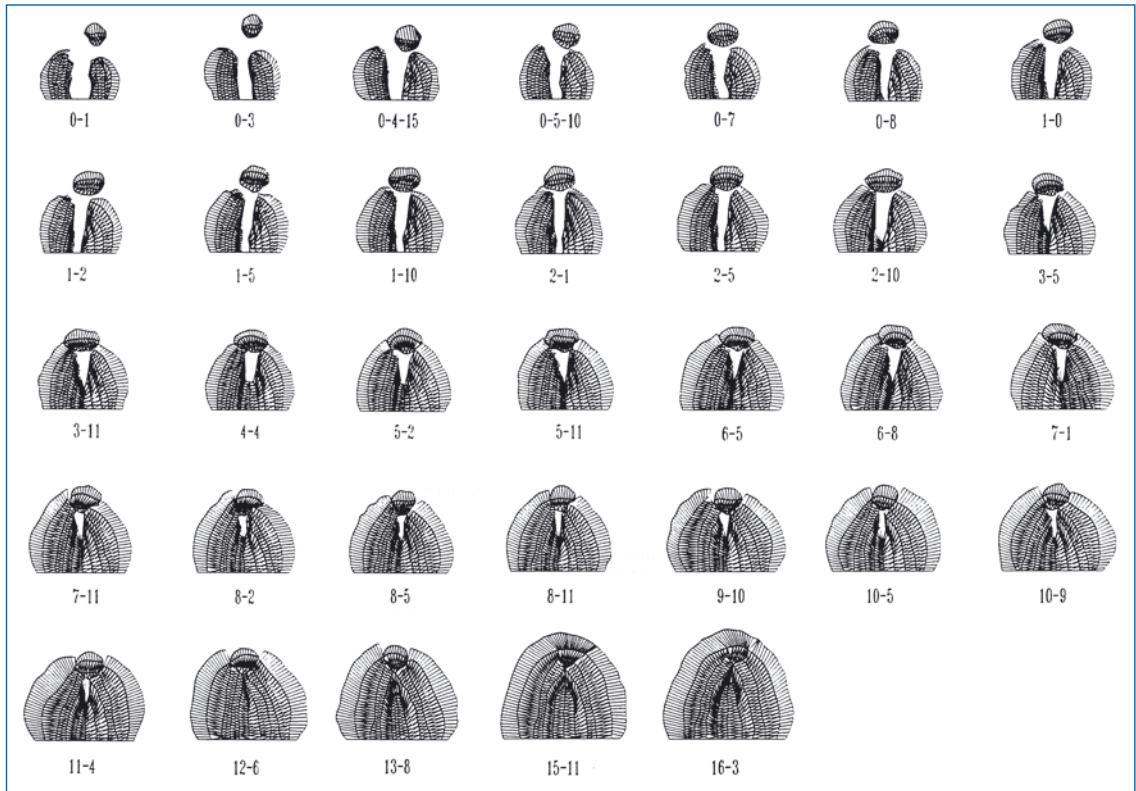


Fig. 6C.25. Case DK (AI-31). Computerized drawings of palatal casts using an electromechanical digitizer. Each cast form is drawn to scale but not its size. The premaxilla is gradually aligned within the lateral palatal segments after the initial ventroflexion. The ratio of palatal size to cleft size increases

with growth. The anterior cleft space is left open until a secondary alveolar bone graft is placed. The palatal cleft was closed at 4-0 leaving an anterior palatal fistula which was closed with a secondary alveolar bone graft at 12-1

Table 6C.2. Surface Area of CBCLP, Case DK (AI-31).

Age	Skeletal Area				Cleft Space			Total
	Premax	RLS	LLS	Tot	Ant	Post	Tot	SA + CS
0-1	55.6	314.1	254.2	623.9	102.9	138.8	341.7	965.6
0-3	79.2	366.6	317.5	763.3	150.5	191.8	342.4	1105.7
0-4-15	86.1	362.4	300.8	749.3	56.1	259.3	315.4	1064.7
0-5-10	99.5	356.2	322.5	778.2	70.3	175.8	245.5	1023.7
0-7	111.7	361.9	321.3	794.9	45.2	168.0	213.2	1008.1
0-8	106.8	376.0	351.0	833.8	85.6	149.1	234.7	1068.5
1-0	100.3	428.1	327.2	855.6	55.6	198.8	249.4	1105.0
1-2	132.8	432.6	389.3	954.7	55.7	189.8	245.5	1200.2
1-5	127.6	507.3	418.1	1053.0	51.2	180.8	232.0	1285.0
1-10	132.6	493.4	421.1	1047.1	39.4	194.9	234.3	1281.4
2-1	117.8	492.6	473.0	1083.4	32.3	168.8	201.1	1284.5
2-5	116.7	503.0	456.0	1075.7	16.1	177.3	193.4	1269.1
2-10	133.6	556.3	449.3	1139.2	17.8	191.2	209.0	1348.2
3-5	102.2	451.8	434.3	988.3	11.2	97.7	108.9	1097.2
3-11	110.7	506.1	427.6	1044.4		94.4	94.4	1138.8
4-4	112.1	501.9	438.5	1052.5		95.4	95.4	1147.9
5-2	115.3	547.0	481.3	1143.6		97.2	97.2	1240.8
5-11	112.2	541.5	514.0	1167.7		82.5	82.5	1250.2
6-5	106.9	646.8	547.5	1301.2		85.7	85.7	1386.9
6-8	102.2	635.8	552.1	1290.1		97.8	97.8	1387.9
7-1	102.1	638.8	591.7	1332.6		67.8	67.8	1400.4
7-11	107.1	667.2	592.4	1366.7		53.3	53.3	1420.0
8-2	104.8	650.9	624.4	1380.1		40.6	40.6	1420.7
8-5	72.5	647.0	597.4	1316.9		40.5	40.5	1357.3
8-11	68.9	704.2	638.8	1411.9		41.6	41.6	1453.5
9-10	81.7	770.5	708.4	1560.6		42.8	42.8	1603.4
10-5#	64.5	726.4	702.4	1493.3		44.9	44.9	1538.2
10-9	86.5	745.0	704.0	1535.5		55.6	55.6	1591.1
11-4	109.9	782.8	804.1	1696.8		32.2	32.2	1729.0
12-6	103.7	799.8	895.7	1799.2				1799.2
13-8	81.1	895.4	914.3	1890.8				1890.8
15-11	83.8	1060.6	1037.2	2181.6				2181.6
16-3	75.4	1086.4	1069.3	2231.1				2231.1

Note: *Premax* = Premaxilla; *RLS* = Right Lateral Segment; *LLS* = Left Lateral Segment; *Tot* = Total Surface Area; *Ant* = Anterior Cleft Space; *Post* = Posterior Cleft Space; *Tot* = Ant + Post; *SA + SC* = Bony Surface Area + Cleft Space Area; #: Changing teeth.

Fig. 6C.26 a,b. Case DK (AI-31). **a, b** Serial lateral cephaloradiographic tracings show changes in the skeletal and soft tissue profile. The upper lip gradually became recessive, reflecting the lack of midfacial growth and superimposed Basion Horizontal facial polygons of Coben. The midfacial protrusion seen at 12 years, 5 months is no different to that seen at 5 years, 5 months of age. At 10 years of age, the midface was more recessive, but with the application of premaxilla advancement orthodontics for 2 years, it was positioned more anteriorly. The lower jaw showed progressive downward and forward growth. As in all faces that grow well, this growth pattern was mainly responsible for the flattening of the facial profile. Comments: I must stress that the Coben analysis demonstrates the forward/downward growth of the entire maxillary and mandibular growth, therefore, the maxilla actually recede due to growth at the upper and lower face

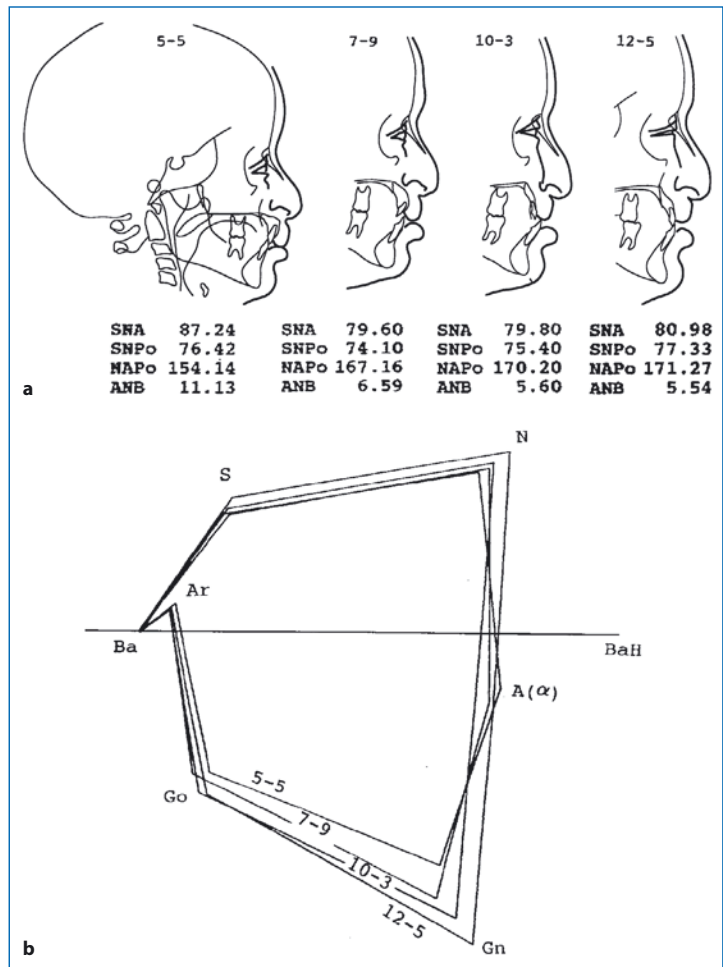




Fig. 6C.27 a–p. Case PM (KK-22). Serial photographs demonstrate excellent facial and palatal growth in CBCLP. Time is an ally! No presurgical orthopedics other than the use of external elastics off a head bonnet for 10 days prior to lip surgery; arm restraints were used to prevent removal of the head bonnet. The palatal cleft was closed at 8 years of age using a modified von Langenbeck procedure. Secondary alveolar cranial bone grafting was done at 8 years of age. **a** and **b** At birth, an extremely

protrusive premaxilla. **c** Head bonnet with elastic over the premaxilla. **d** The lip cleft was closed using Millard's forked flap procedure. The tissue in the nostril is used to reconstruct the columella at a later date so that the surgeon need not go back to the lip for tissue. **e** Occlusal view at 5 years of age. Note that the premaxilla is still protrusive. **f** The lip is being pushed forward by the protruding premaxilla. The palatal cleft was closed at 8–6. The face at 5 years showing upper lip protrusion



Fig. 6C.27 a–p. (continued) **i** 11-01 The premaxilla is still protrusive. **j, k, and l** At 20 years of age the occlusion is ideal. Minor imperfections in the anterior teeth were corrected by the use of composite material. **m** Ideal arch form with a normal palatal

vault space. Good alveolar bone support to the lateral incisors. **n, o, and p** Facial photographs at 20 years of age showing a harmonious and pleasing soft tissue profile. Comments: No protraction midfacial mechanics were used

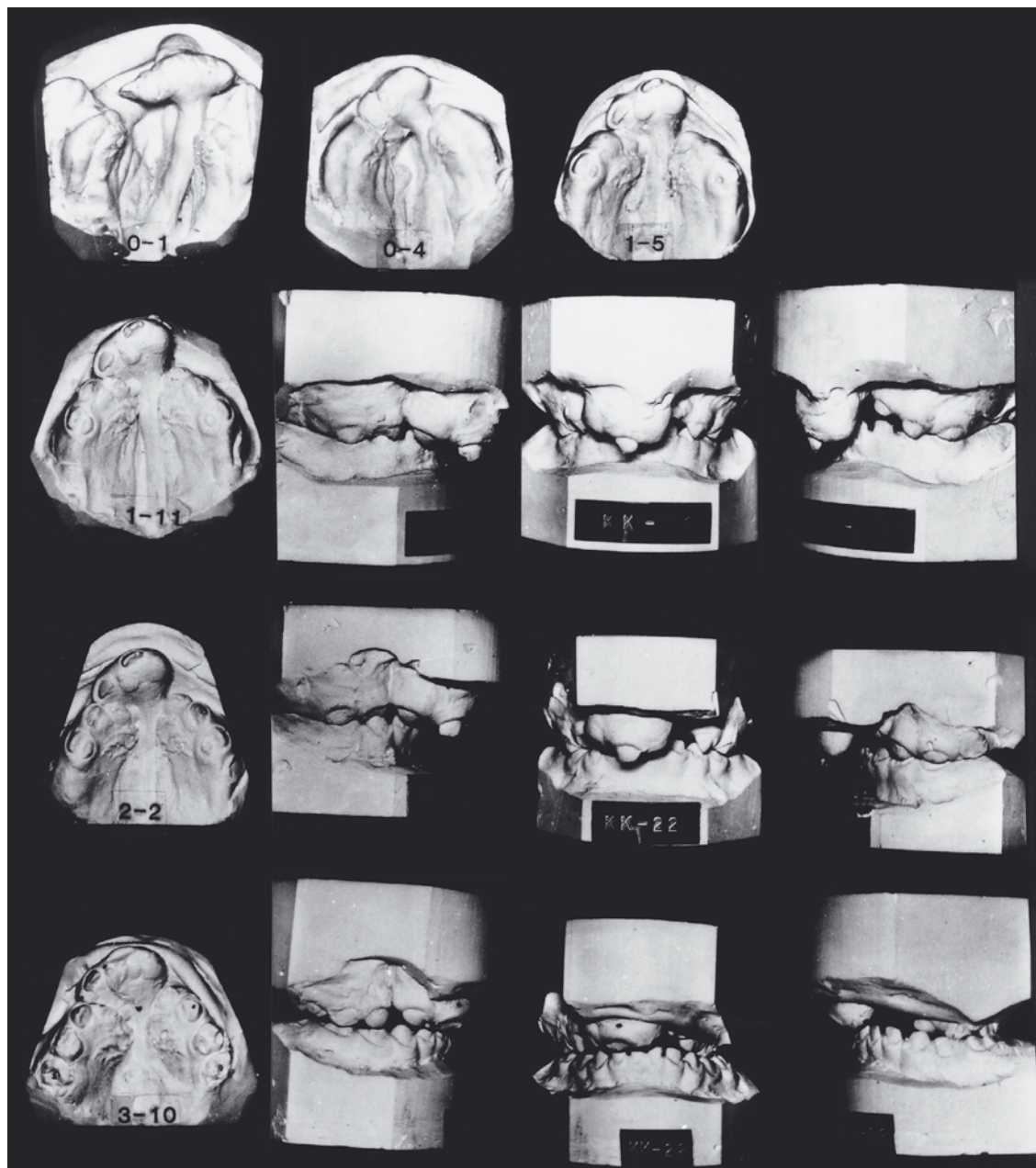


Fig. 6C.28. Case PM (KK-22). Serial casts: 0-1 At birth the septum is deviated to the left, and the right palatal segment is laterally displaced. 0-4 With an external elastic and lip surgery, the premaxilla flexes ventrally and medially, making contact

with both palatal segments. The remaining casts show excellent buccal occlusion in Class II relationship with overbite and severe anterior overbite and overjet

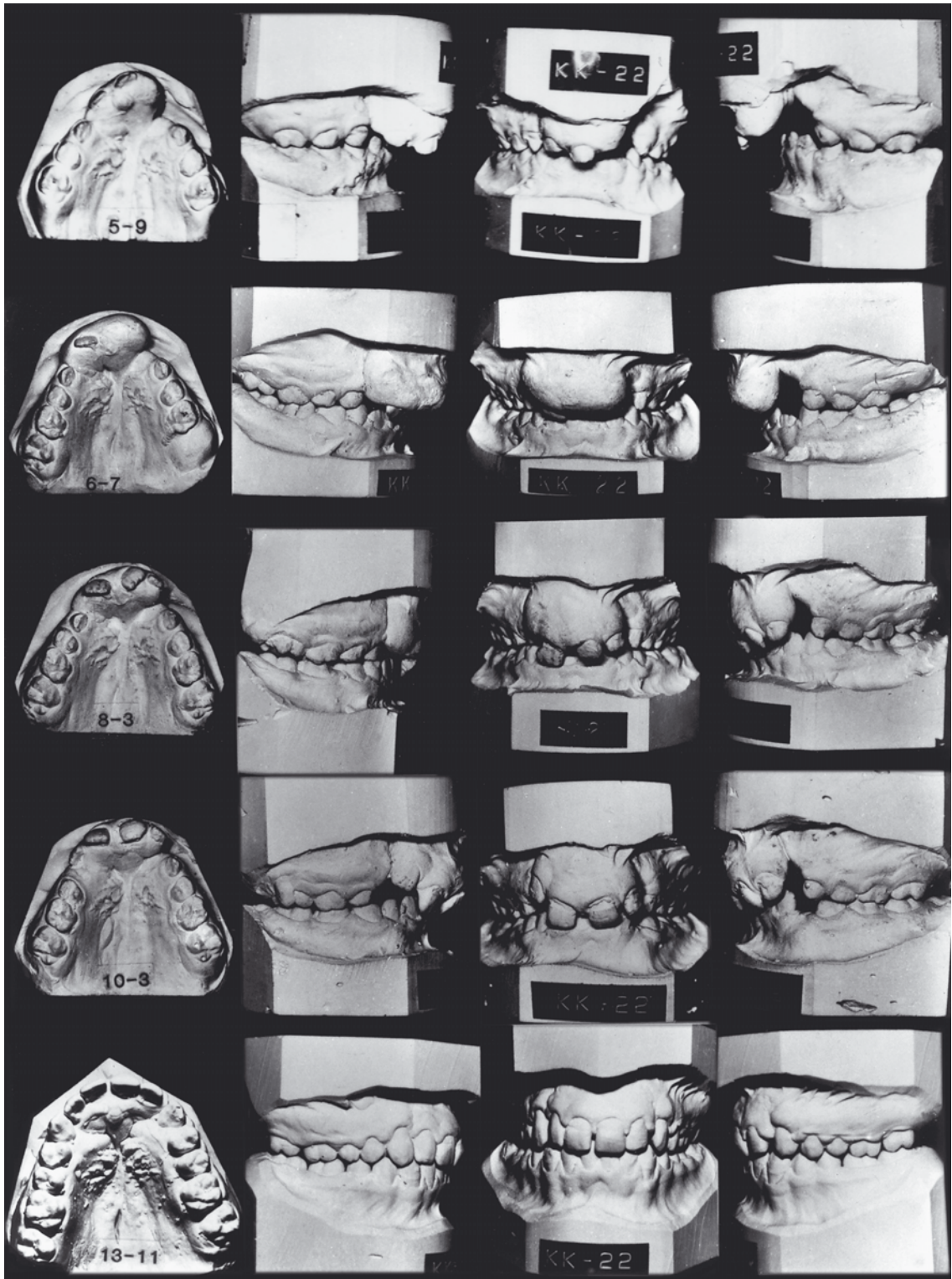


Fig. 6C.28. (continued) By 8-3 the anterior overjet is markedly reduced by growth. 13-11 Conventional orthodontics were eventually used to reduce the Class II occlusion and align the anterior teeth into an ideal overbite-overjet relationship

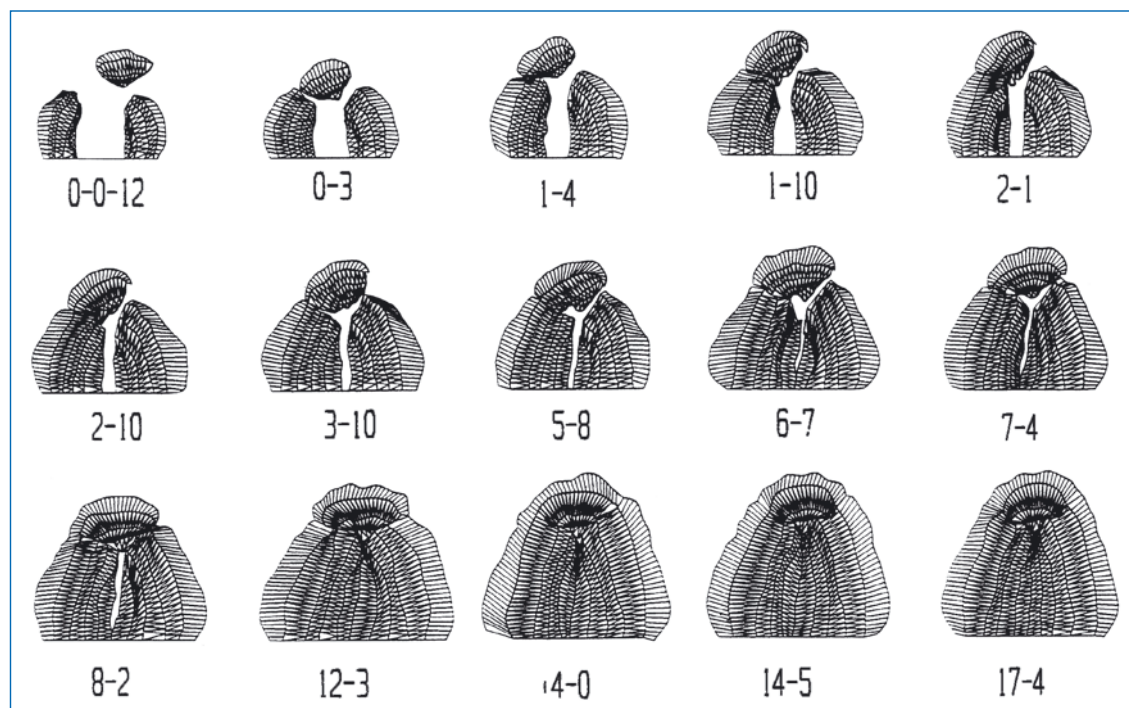


Fig. 6C.29. Case PM (KK-22). Computer-generated outlines of the serial casts were performed using an electromechanical digitizer. All casts are drawn to scale. The casts range from 12 days to 17 years, 4 months of age. This series demonstrates the spontaneous closure of the anterior and posterior cleft spaces after

“molding action” brought on by uniting the lip and then by gradual palatal growth at the border of the cleft space. The premaxilla was initially aligned forward of the lateral palatal segments but was satisfactorily incorporated within the arch at a later age

Table 6C.3. Surface Area of CBCLP. Case PM (KK-22). The palatal surface area increased by 4096 after 1 year, 4 months and by 7696 at 2 years, 1 month. By 8 years, 2 months, the palatal surface area had increased two and a half times rohen the cleft space was closed.

Age	Skeletal Area				Cleft Space			Total
	Premax	RLS	LLS	Tot	Ant	Post	Tot	SA + CS
0-0-12	145.5	335.5	282.7	763.7	127.4	417.0	544.4	1308.1
0-3	150.2	397.4	377.6	925.2	65.4	331.5	396.9	1322.1
1-4	154.0	469.5	464.3	1087.8	36.8	265.5	302.3	1390.1
1-10	211.8	502.6	506.2	1220.6	70.4	216.3	286.7	1507.3
2-1	217.6	589.0	549.5	1356.1	79.8	195.3	275.1	1631.2
2-10	220.7	603.9	551.3	1375.9	95.8	193.2	289.0	1664.9
3-10	271.6	660.4	616.5	1548.5	122.6	206.3	328.9	1877.4
5-8	273.3	673.0	675.9	1622.2	123.6	201.2	324.8	1947.0
6-7	273.6	811.0	820.5	1905.1	115.0	206.5	321.5	2226.6
7-4	277.3	813.0	839.5	1929.8	106.7	185.2	291.9	2221.7
8-2	306.5	844.6	890.8	2041.9	101.1	155.4	256.5	2298.4
12-3	346.8	1087.1	1116.1	2550.0				2550.0
14-0	348.7	1161.8	1226.4	2736.9				2736.9
14-5	351.1	1198.8	1237.0	2786.9				2786.9
17-4	353.5	1241.0	1246.3	2840.8				2840.8

Note: *Premax* = Premaxilla; *RLS* = Right Lateral Segment; *LLS* = Left Lateral Segment; *Tot* = Total Surface Area; *Ant* = Anterior Cleft Space; *Post* = Posterior Cleft Space; *Tot* = Ant + Post; *SA + SC* = Bony Surface Area + Cleft Space Area; #: Changing teeth.

Fig. 6C.30. Case PM (KK-22). Palatal outlines were superimposed using the rugae for registration. This series shows that the premaxilla's position within the maxillary complex at 17 years of age is similar to that seen at birth. Excellent growth occurs in all dimensions and is similar to the growth pattern seen in noncleft palates. Increased posterior palatal growth is necessary to accommodate the developing molars. Alveolar bone growth with tooth eruption increases midfacial height. Comments: The position of the anterior premaxilla relative to the anterior cranial base (Nasion) to the anterior position of pogonion of the mandibular symposium shows the same relative position from birth to 17 years of age. These 2 studies confirm that midfacial growth is retarded

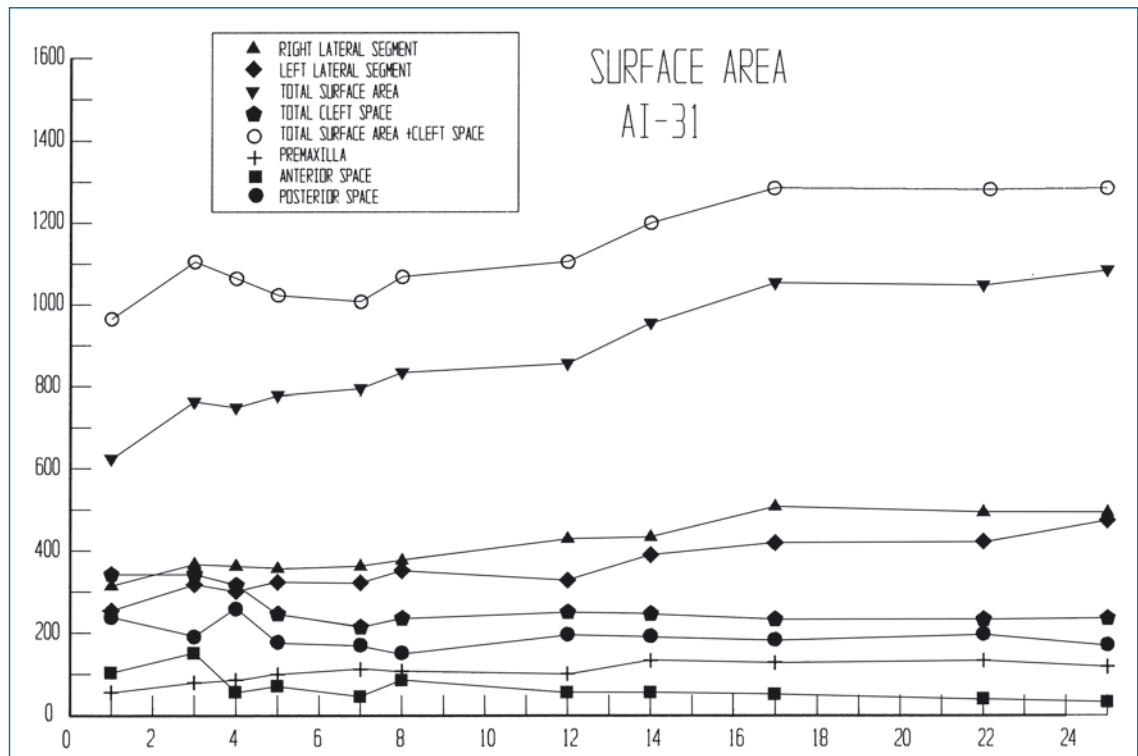
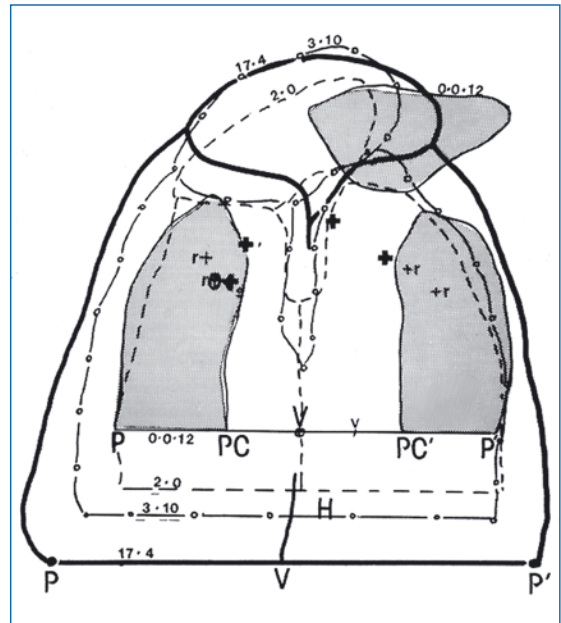


Fig. 6C.31. Case DK (AI-31). Premaxillary ventroflexion with medial movement of the lateral palatal segments caused a great reduction in the anterior and posterior cleft spaces by 5 months of age. Thereafter, for the next 19 months, the anterior cleft

space gradually reduced, while the posterior cleft space showed some increase due to the increase in palatal length. Both lateral palatal segments showed a similar, gradually increasing growth rate

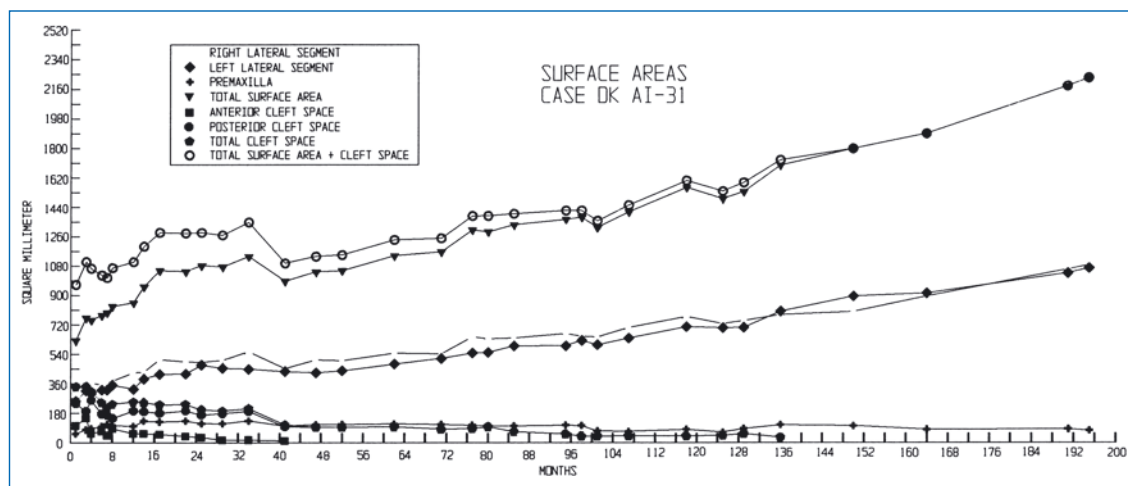


Fig. 6C.32. Case DK (AI-31). After the initial change in cleft size brought on by medial movement of the lateral palatal segments and ventroflexion of the premaxilla, the greatest acceleration of cleft space closure occurred between 2-10 and 3-5. The premaxilla reached its largest size by 3 years, which is associated with eruption of the teeth. Palatal growth acceleration oc-

curred between 1 to 3 months and 12 to 14 months and then gradually tapered off. The palatal segments had increased 37% in size by 1 year and 74% by 2 years. Palatal growth at its medial borders still occurs; even though it narrows the cleft space, the total cleft space is increasing in size due to the increase in palatal length

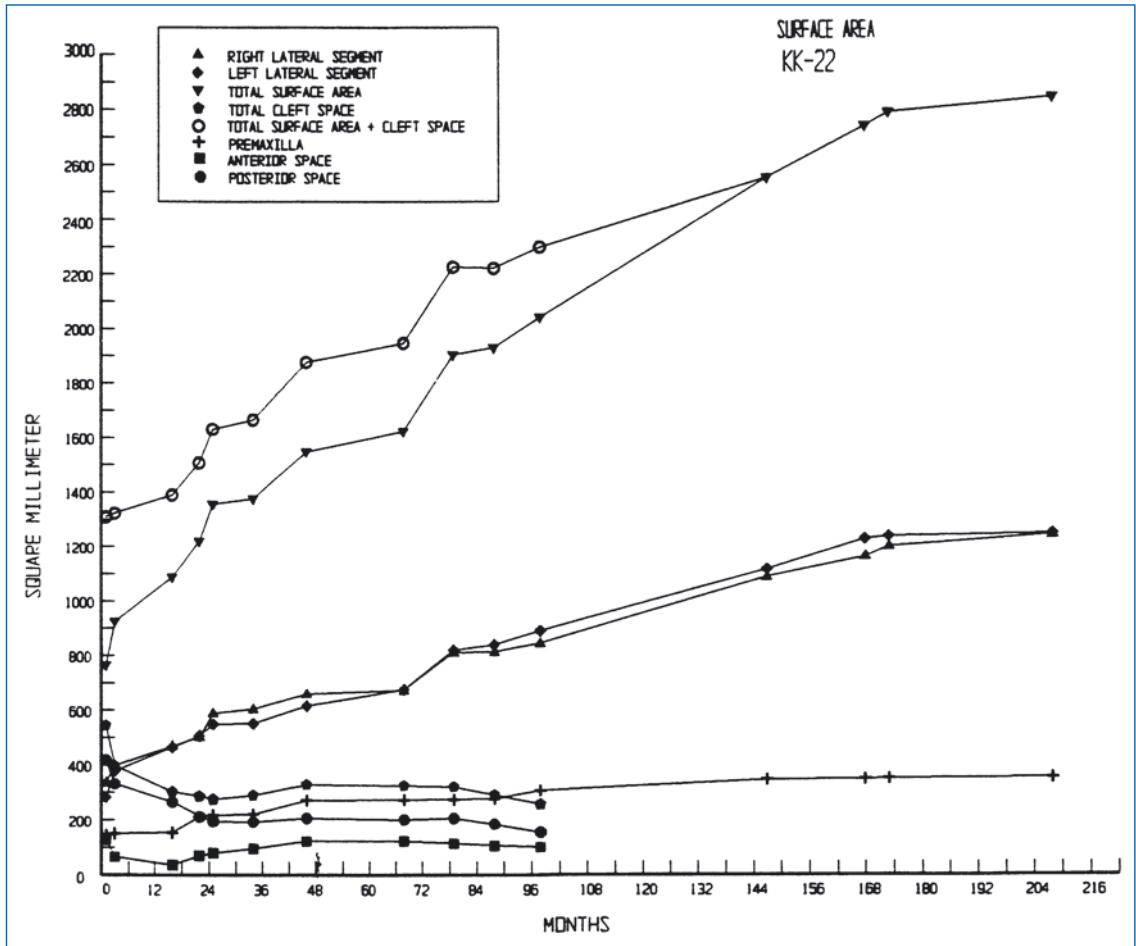


Fig. 6C.33. Case PM (KK-22) Time sequence analysis of serial palatal growth shows that both palatal segments are growing at the same rate and to the same degree. The premaxilla is also increasing in size with tooth eruption but at a lesser rate. The greatest palatal growth acceleration occurs the first 2 years and then tapers off. The anterior cleft space is initially reduced as a result of premaxillary ventroflexion, but thereafter it remains

the same dimensions until the palatal cleft is closed. The posterior cleft space initially is reduced with palatal medial movement. The resulting posterior cleft space remains approximately the same size for the next 8 years. It must be remembered that the cleft length is increasing while the cleft width is decreasing. The net cleft area is gradually reducing with growth. All fistulae are closed by 12–3 years of age

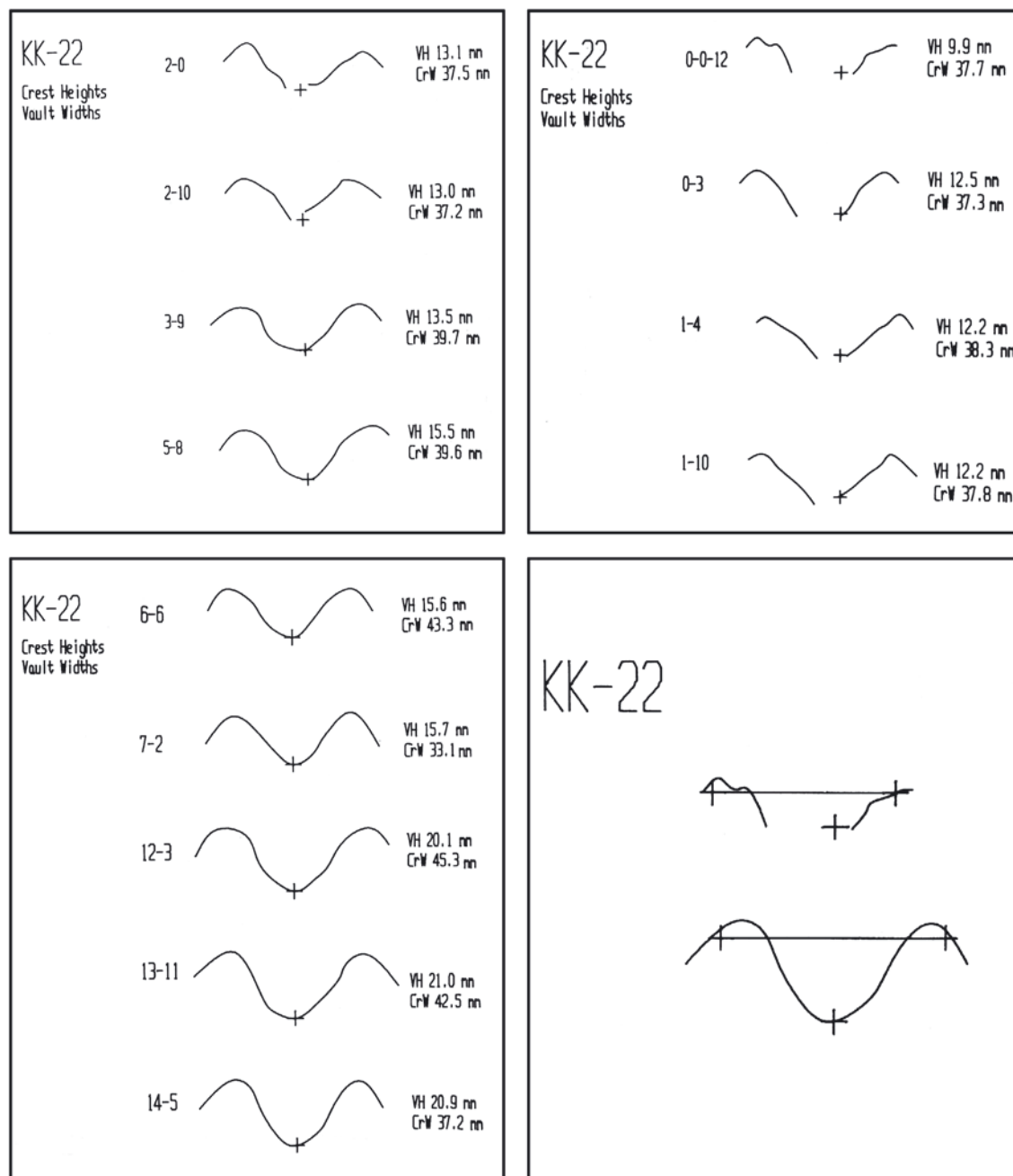


Fig. 6C.34. Serial midpalatal cross-sections showing vault height and palatal width changes using stereophotogrammetry. The left lateral palatal segment is attached to the vomer – while the right lateral palatal segment is displaced laterally. A lip muscle adhesion causes the displaced palatal segment to move medially, narrowing the cleft space. The appositional growth of the alveolar segments is continuous and becomes more obtuse.

Closure of the palatal cleft space with a modified vomer flap maintains a normal vault height, and vault space flattening of the vault occurs in almost all cases in the absence of a vomer flap. Comments: A vomer flap with a von Langenbeck procedure seems to create a minimum scarring. Vomer flap alone performed early (6 months to 1 year) created excessive scarring

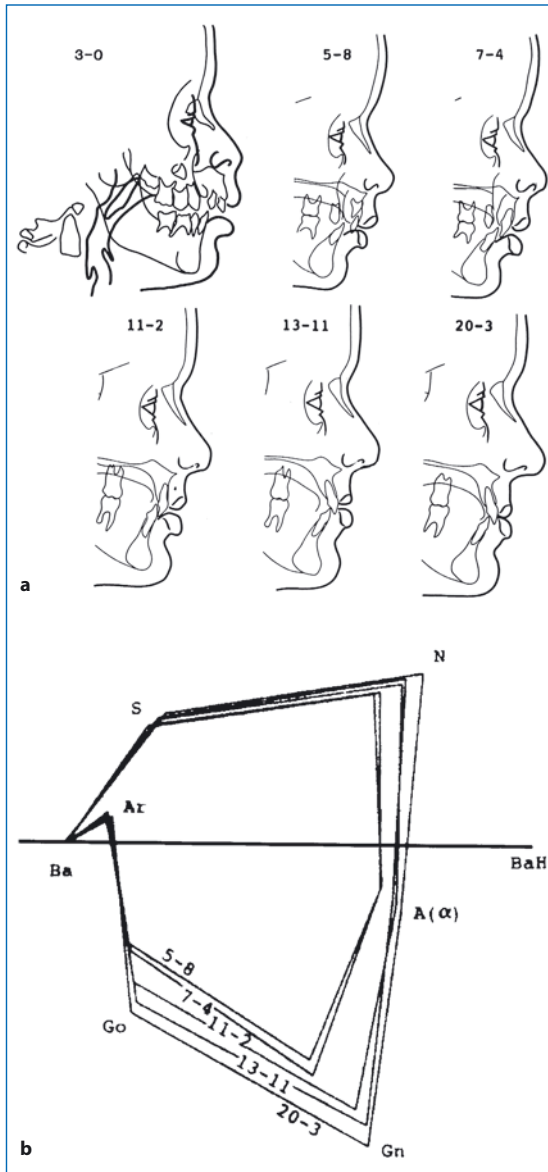


Fig. 6C.35 a,b. Case PM (KK-22) **a** Cephalometric serial tracings of the skeletal and soft tissue profile show marked reduction of the midfacial protrusion. **b** Superimposed serial tracings using Coben's Basion Horizontal method show an excellent facial growth pattern which straightens the skeletal profile. There is very little forward midfacial growth between 11 and 20 years of age. During the same time period, growth at the anterior cranial base and the mandible contributed to flattening of the facial profile

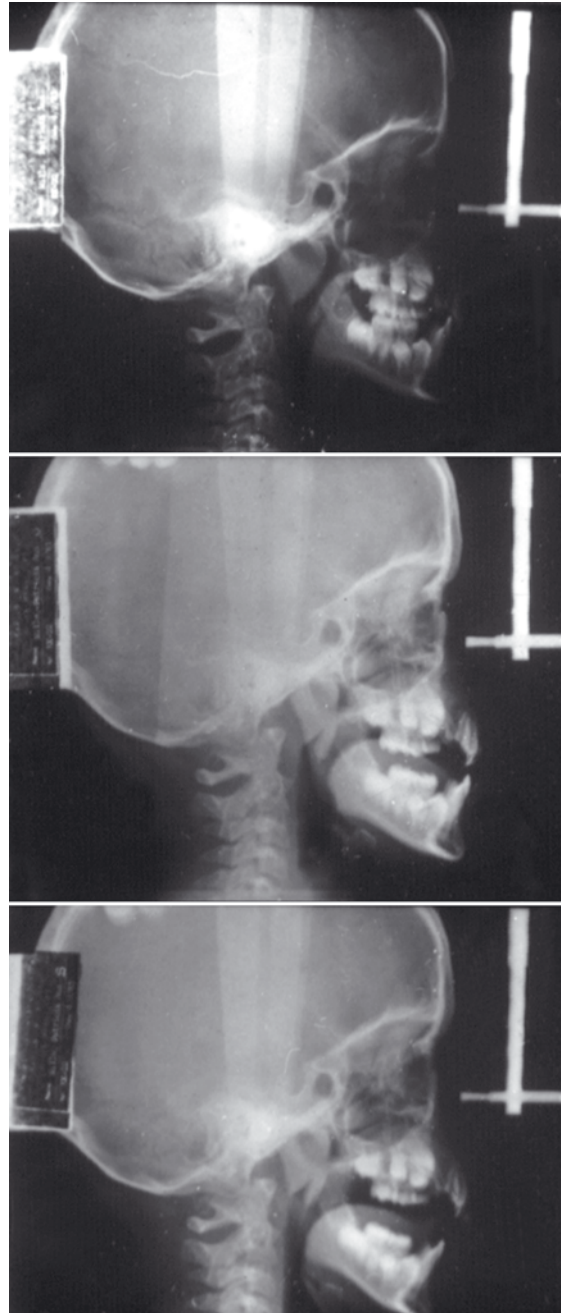


Fig. 6C.36. Case PM (KK-22). Cephaloradiographs taken at 5 years of age. *Top:* At rest with teeth together. *Middle:* Taken while vocalizing "Youu . . ." *Bottom:* Taken while vocalizing "Sss . . ." Comments: When vocalizing both sounds, the velum elevates and makes contact with the adenoids. The pharyngeal depth is relatively small. The adenoids are of moderate size, the velum is of good length and shows good elevation. This is not a functional test to evaluate velopharyngeal closure but it does show the well-proportioned oral and nasal pharyngeal spaces which are conducive to good velopharyngeal closure. A gap space more than 5 mm may indicate VPI exist with inadequate lateral pharyngeal wall movements

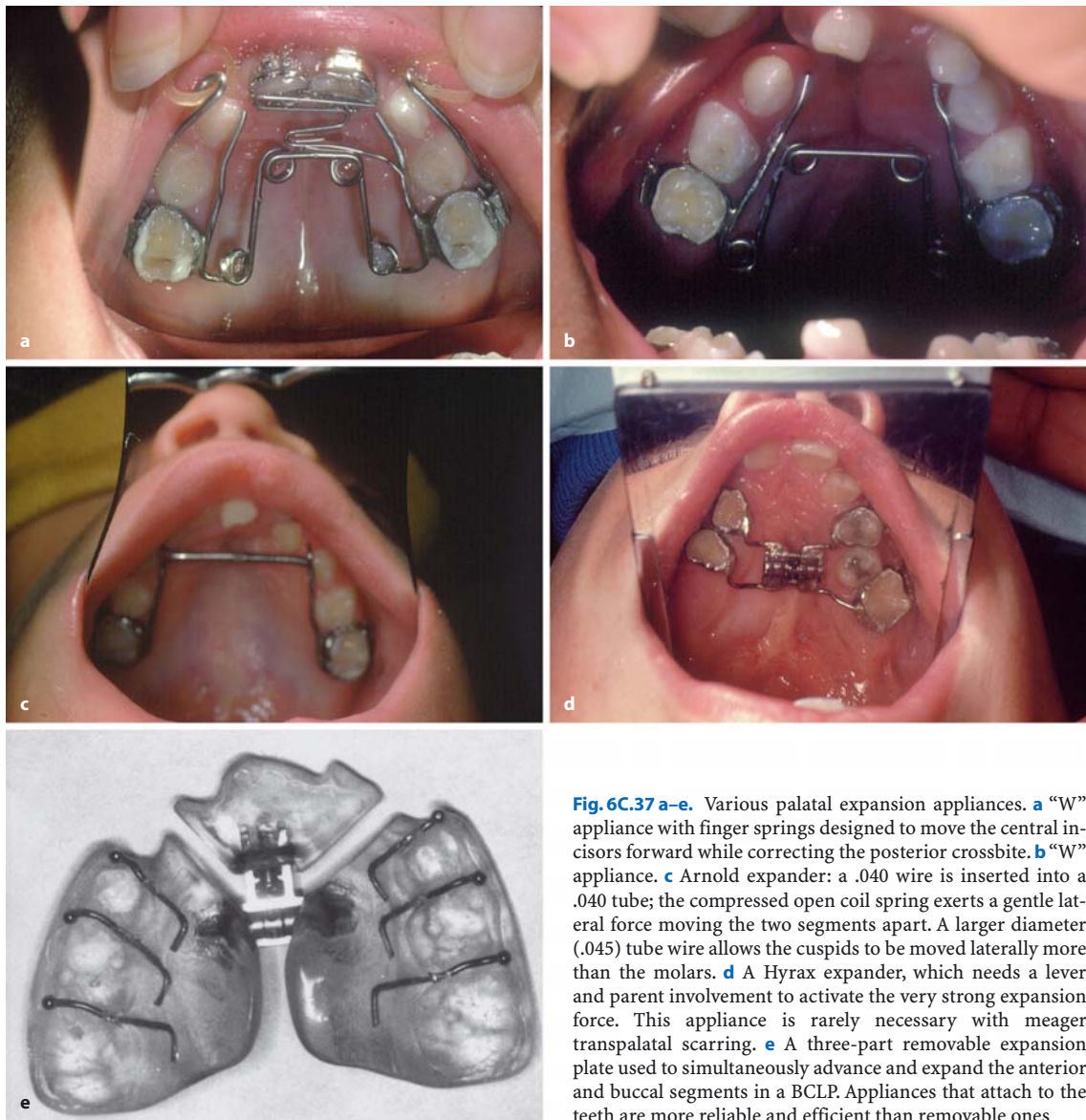


Fig. 6C.37 a–e. Various palatal expansion appliances. **a** “W” appliance with finger springs designed to move the central incisors forward while correcting the posterior crossbite. **b** “W” appliance. **c** Arnold expander: a .040 wire is inserted into a .040 tube; the compressed open coil spring exerts a gentle lateral force moving the two segments apart. A larger diameter (.045) tube wire allows the cuspids to be moved laterally more than the molars. **d** A Hyrax expander, which needs a lever and parent involvement to activate the very strong expansion force. This appliance is rarely necessary with meager transpalatal scarring. **e** A three-part removable expansion plate used to simultaneously advance and expand the anterior and buccal segments in a BCLP. Appliances that attach to the teeth are more reliable and efficient than removable ones



Fig. 6C.38 a–s. Case ML (KK-56) demonstrates severe premaxillary protrusion at birth in IBCLP. “Whisker” forked flap was performed at 2 months, definitive lip surgery at 6 months, and palatal cleft closure at 18 months. Secondary alveolar cranial

bone grafting was placed at 8 years, 3 months. Maxillary surgery with chin augmentation was performed at 15 years, 7 months. **a–g** Facial and intraoral photographs show progressive facial and occlusal changes

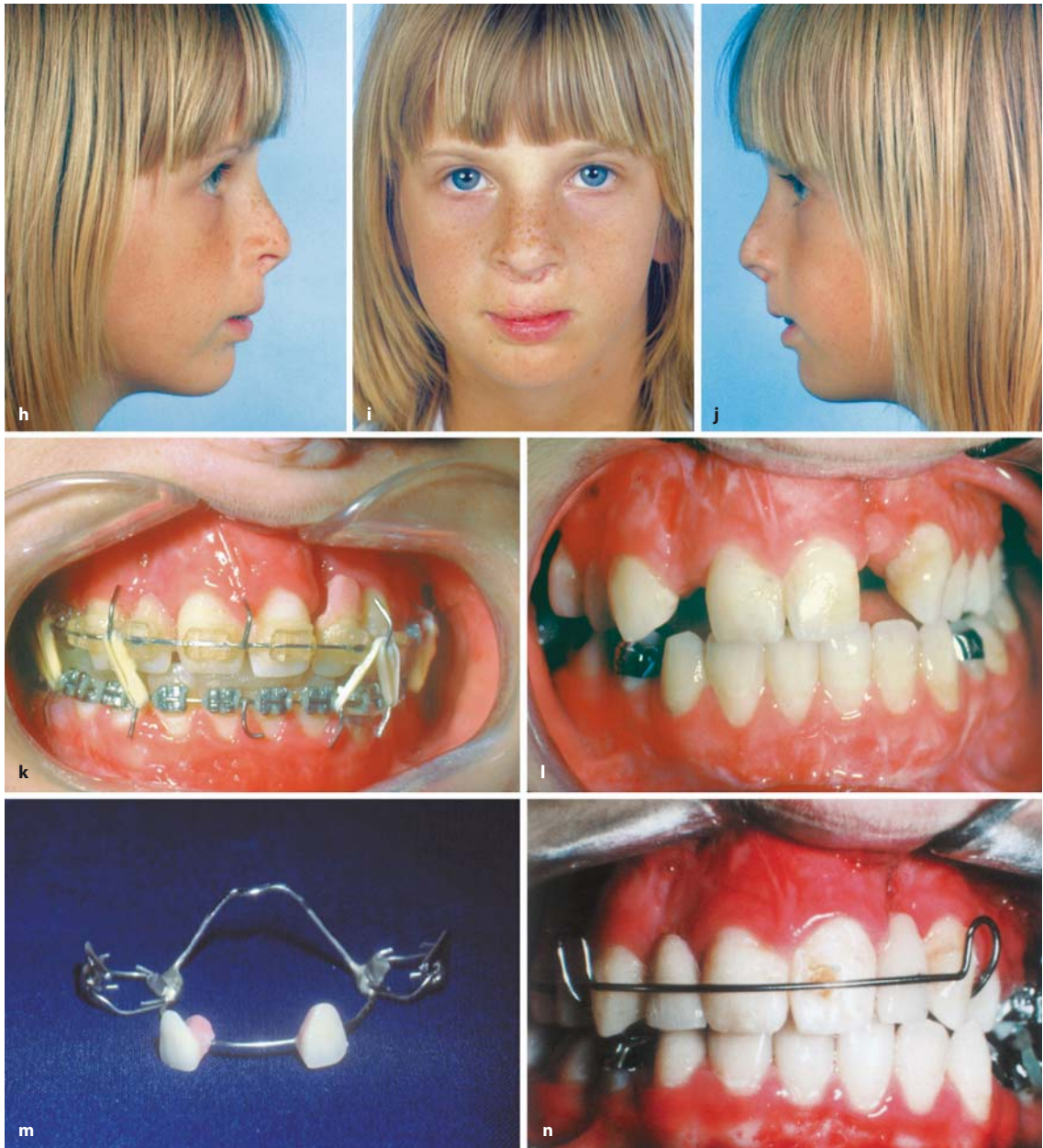


Fig. 6C.38 a-s. (continued) **h, i, and j** Facial photographs at 8 years of age. **k** After Lefort I advancement, the lateral incisor pontics were attached to the arch wire for aesthetics. **l** and **m** Occlusal photographs showing missing incisor spaces



Fig. 6C.38 a-s. (continued) **n**, **o**, and **p** Intraoral photographs showing retainer with lateral incisor pontics in place. **q**, **r**, and **s** Facial photographs at 17 years of age. Case ML (KK-56) demonstrates severe premaxillary protrusion at birth in IBCLP. **h**, **i**, and **j** Facial photographs at 8 years of age. **k** After Lefort I advancement, the lateral incisor pontics were attached to the arch wire for aesthetics. **l** and **m** Occlusal photographs showing missing incisor spaces. **n** A partial upper retainer in place with

pontics for missing lateral incisors. Case ML (KK-56) demonstrates severe premaxillary protrusion at birth in IBCLP. **o**, and **p** Intraoral photographs showing a fixed bridge to replace both upper lateral incisors. **q**, **r**, and **s** Facial photographs at 17 years of age. A prominent symphysis is noted. Comment: This case shows the need to keep the lateral incisors spaces open in order to obtain good anterior overbite and overjet in the presence of strong mandibular growth

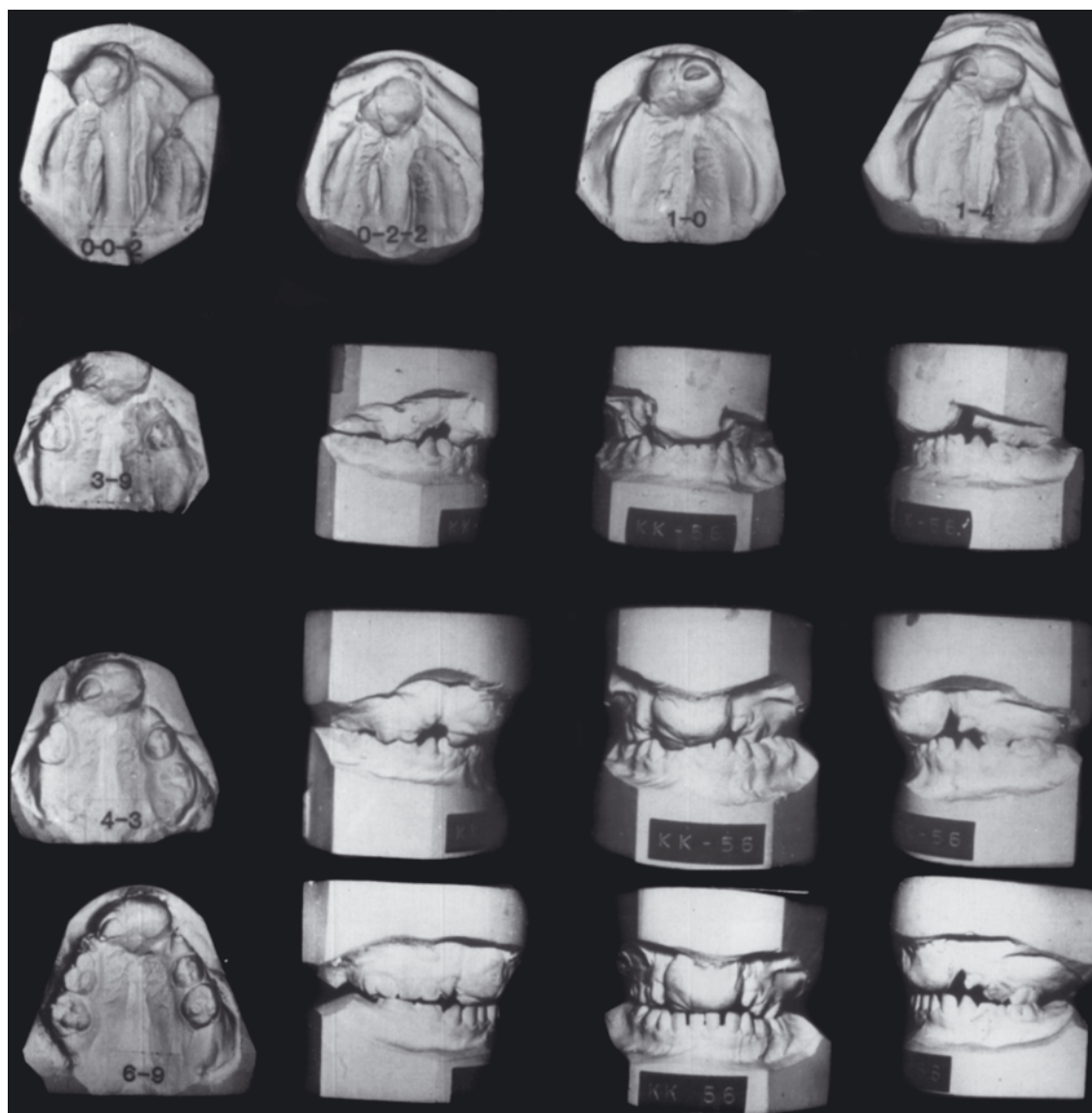


Fig. 6C.39. Case ML (KK-56). Serial casts from 0-0-2 to 4-3: With the establishment of an intact lip musculature, the premaxilla and lateral palatal segments molded into a good arch form. The premaxilla, although latero- and ventroflexed, still

caused the upper lip to be pushed forward. The left buccal crossbite was corrected by 8 years of age and a fixed palatal retainer placed

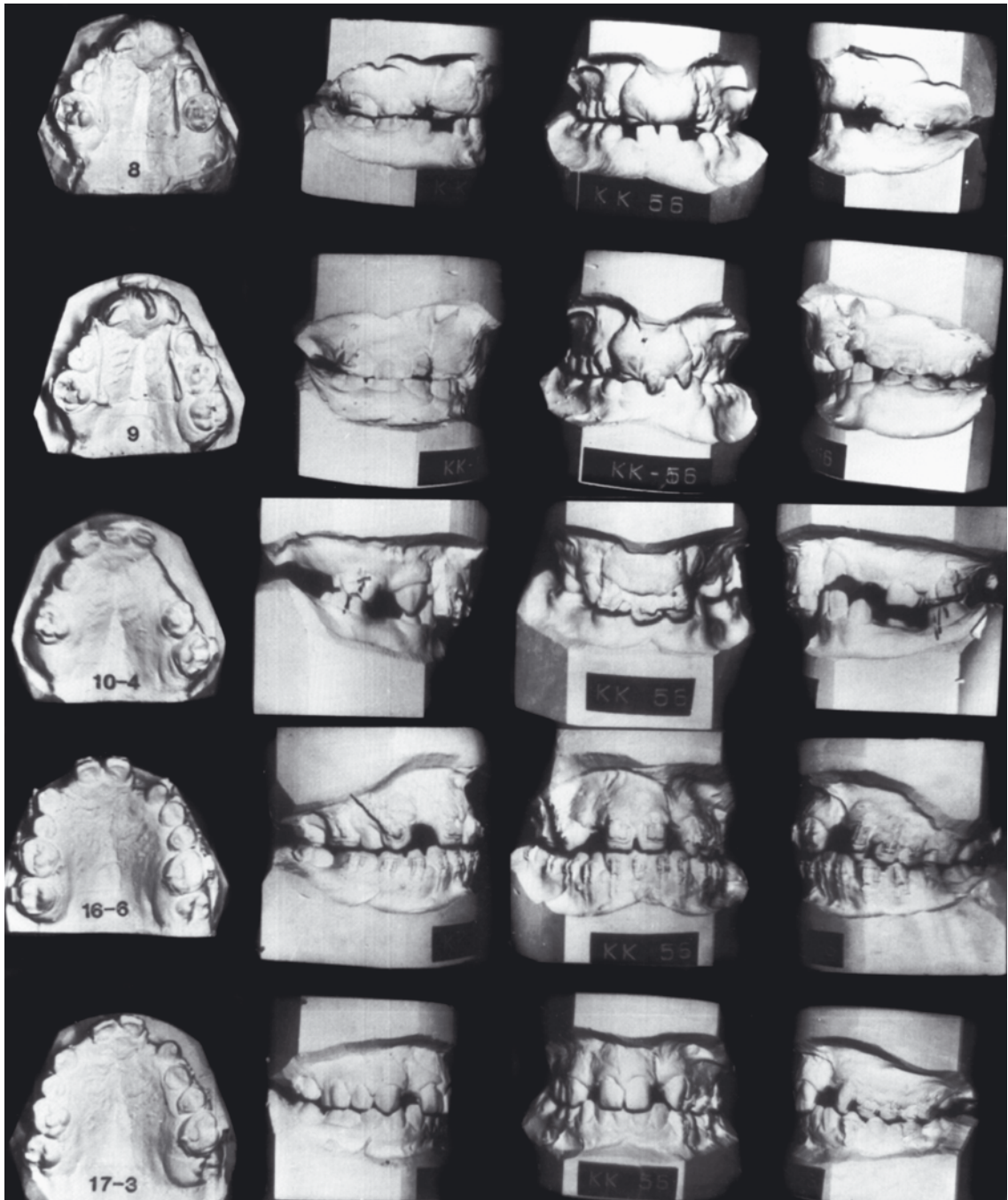


Fig. 6C.39. (continued) 15-7 Orthodontic treatment was designed not to correct the slight Class II occlusion of the left segment. 15-9 After Lefort I osteotomy and final teeth alignment. Because the premaxilla was positioned slightly to the right and could not be centered orthodontically, it was decided to leave

the left occlusion in Class II and the right occlusion in Class I, thereby equalizing the space for the lateral incisors. 17-0 A cuspid-to-cuspid fixed bridge replaced the missing lateral incisors and stabilized the relationship of all segments

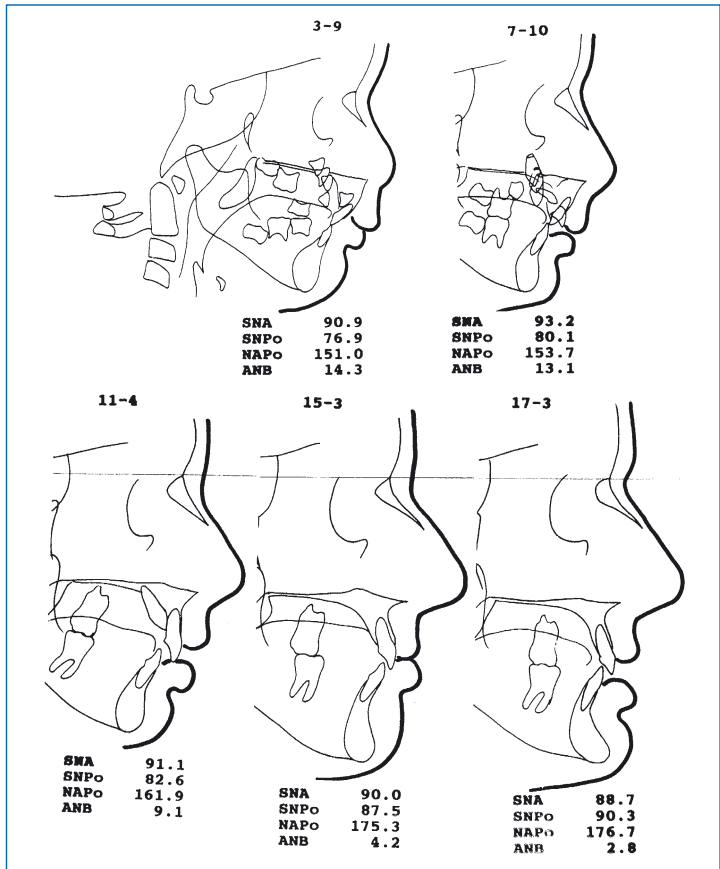


Fig. 6C.40. Case ML (KK-56). Serial cephalometric tracings showing well-proportioned facial growth with a flattening of the facial profile. The protrusive premaxilla was present at 7-10. Orthodontia at 11 years of age improved the axial inclination of the maxillary incisors. The profile at 15-3 is more attractive than that at 17-3 as a result of the chin augmentation at 15-7. The chin point is too protrusive, resulting in a prominent sublabial fold

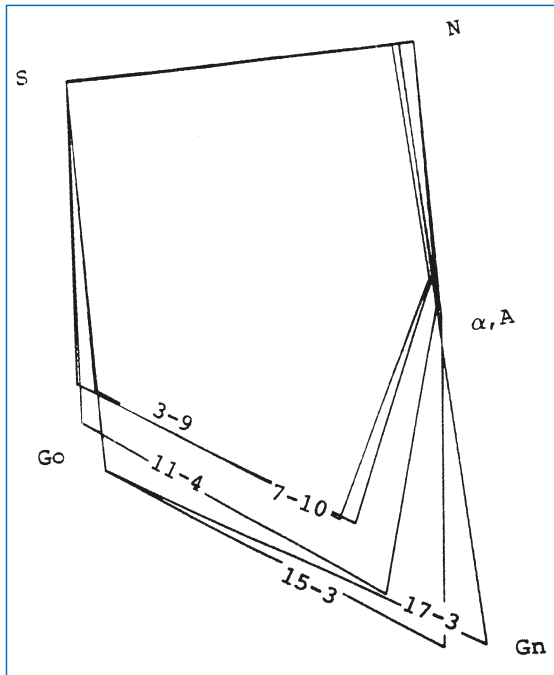


Fig. 6C.41. Case ML (KK-56). Superimposed polygons show an excellent facial growth pattern flattened the profile by 15-3. Midfacial osteotomy corrected the maxillary asymmetry. A chin augmentation was performed at 15-7 years which created a too prominent chin. The mandible continued to grow until 17-3 years of age, creating a slightly concave skeletal profile. The patient is considering having the chin prominence reduced. Comment: Rarely should a chin augmentation be performed with a LeFort I advancement to avoid creating a “dished in” face if the midfacial advancement relapses. Note the small forward growth increments at the anterior cranial base and midface. The midfacial changes did show good vertical growth to maintain normal facial proportions

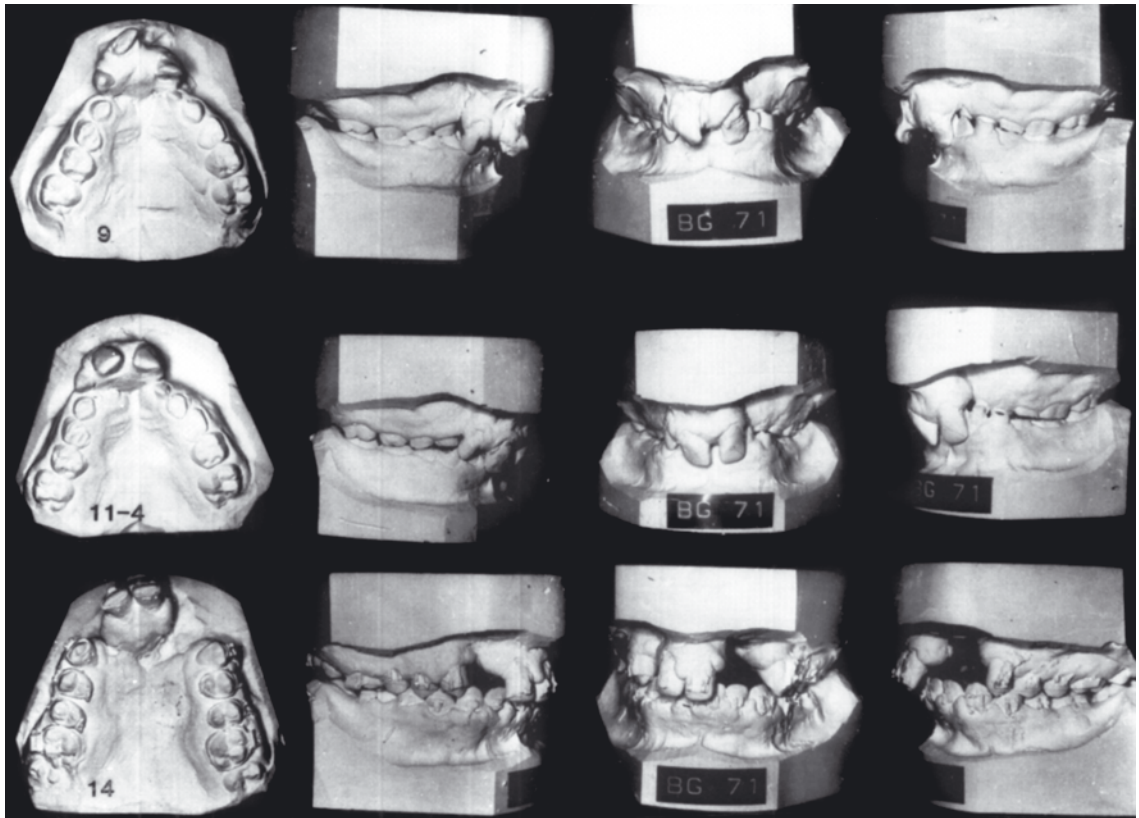


Fig. 6C.42. Case CW (BG-71). Serial casts demonstrate forward advancement of the buccal segments to reduce a very large anterior palatal cleft space. There are instances when a marked osteogenic deficiency exists in both lateral incisor areas where the premaxilla is in a slight overjet-overbite relationship and the buccal segments in a good Class I relationship. In these cases, there may be insufficient contiguous soft tissue to close the anterior palatal cleft space when performing a secondary alveolar bone graft and create a normal site for tooth replacement. The

treatment of choice is to advance both buccal segments, simultaneously placing secondary alveolar bone grafts, yet leaving space for the lateral incisors. After surgery the cuspids were to return to their original Class I position. It was believed that with early premaxillary setback there would be inadequate soft tissue to obtain adequate cleft closure, but worst of all it would have created a severely retruded midface which would have required midfacial advancement

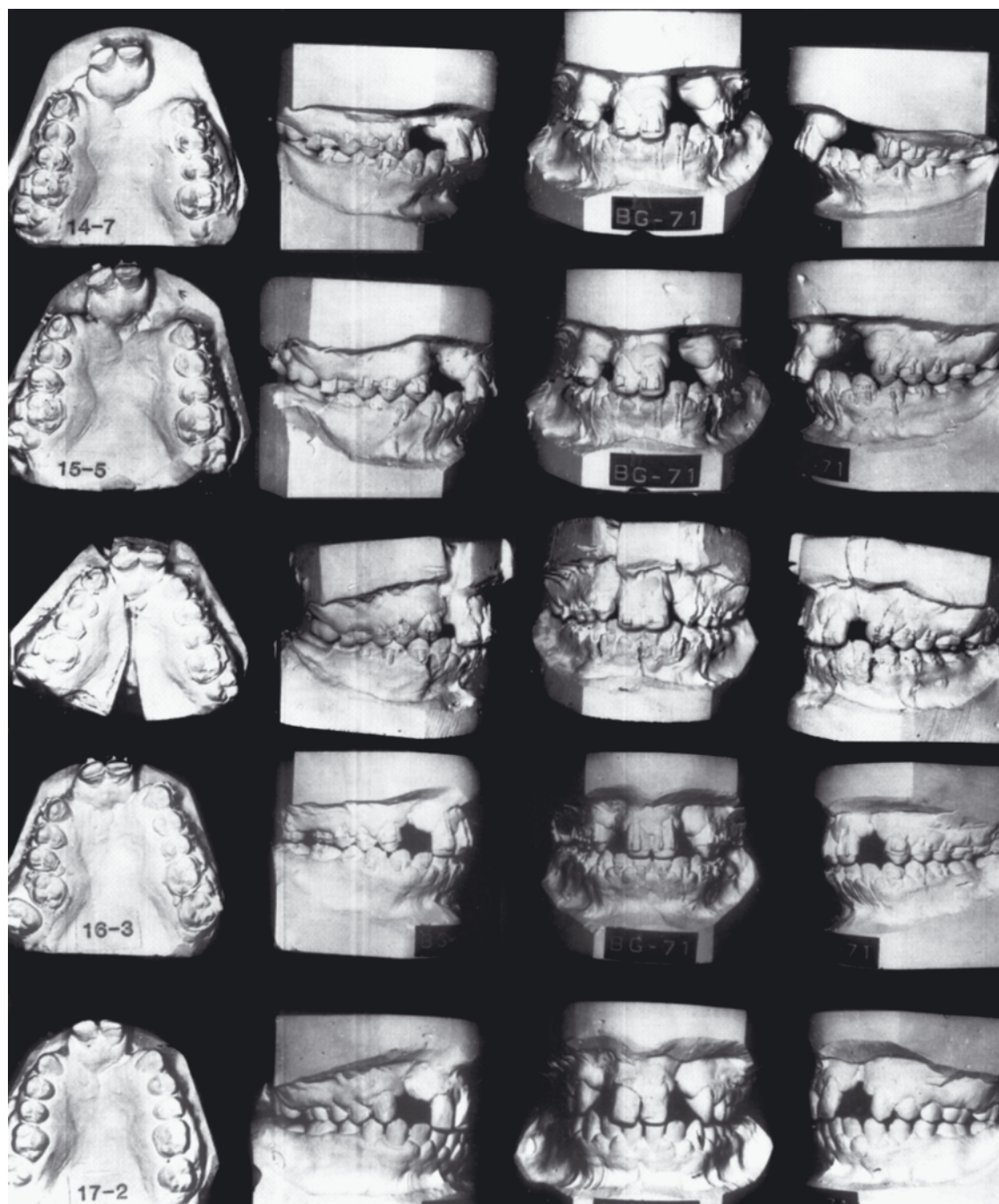


Fig. 6C.42. (continued) 15-5 Good premaxillary relationship with a large anterior palatal cleft space. Class I posterior occlusion. Sectioned plaster casts with the posterior segments placed in a Class II relationship. 16-3 and 17-2 Both buccal segments relapsed into Class I. The main objective of closing the anterior palatal cleft spaces was achieved. Final casts show a good Class

I occlusion with a satisfactory overjet-overbite relationship. The anterior palatal cleft space was closed. However, the alveolar bone graft did not take. In most cases the advanced lateral palatal segments will remain forward with the cuspids in the lateral space



Fig. 6C.43 a-z. Case CS (AF-48) demonstrates severe premaxillary protrusion in a child with CBCLP at birth, resulting in maxillary retrusion in adolescence with the eventual loss of the premaxillary incisors. Lip adhesion was performed at 3 months with forked flap and posterior palate cleft closure at 3 years. No secondary alveolar bone grafts were utilized. Premaxillary sur-

gical advancement at 15 years to correct its retrusion. The premaxillary incisors were extracted due to severe periodontal bone loss, and the anterior palatal oronasal opening was closed at 16 years of age. **a** and **b** Newborn. **c-i** Even with premaxillary ventroflexion, the upper lip was still pushed forward

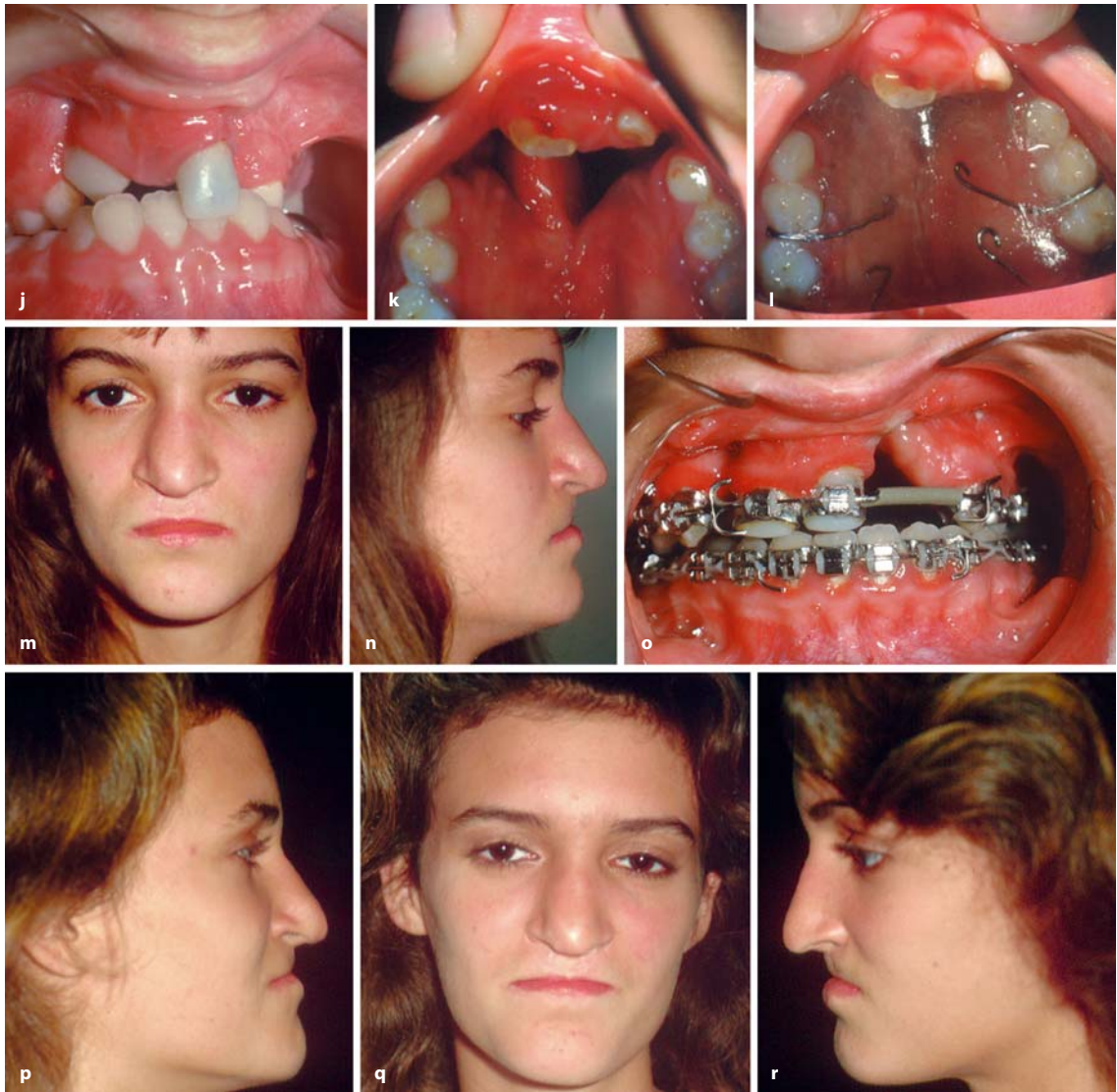


Fig. 6C.43 a-z. (continued) **i-l** A plastic obturator was utilized at 6 years of age to close the very large anterior palatal cleft space to aid speech development and feeding. **m-o** At 14 years,

a retrusive looking midface with an extremely tight upper lip and depressed nasal tip masked a good premaxillary overjet



Fig. 6C.43 a-z. (continued) **u** At 15 years following an unsuccessful attempt to surgically center the premaxilla and close the anterior cleft space. The maxillary incisor roots began to show severe external root absorption. A very large anterior cleft space remains. **v-y** After lip and nose revision, the anterior teeth were extracted and the oronasal opening closed with adjoining soft tissue. **z** A removable maxillary prosthesis replaced missing

teeth and bumpered the upper lip forward. Comments: It would have been better treatment to have surgically advanced both palatal segments to close the very large anterior palatal cleft space. The root absorption was secondary to traumatic orthodontics utilized to maintain the incisor overjet with a protective facial mask and cross elastics

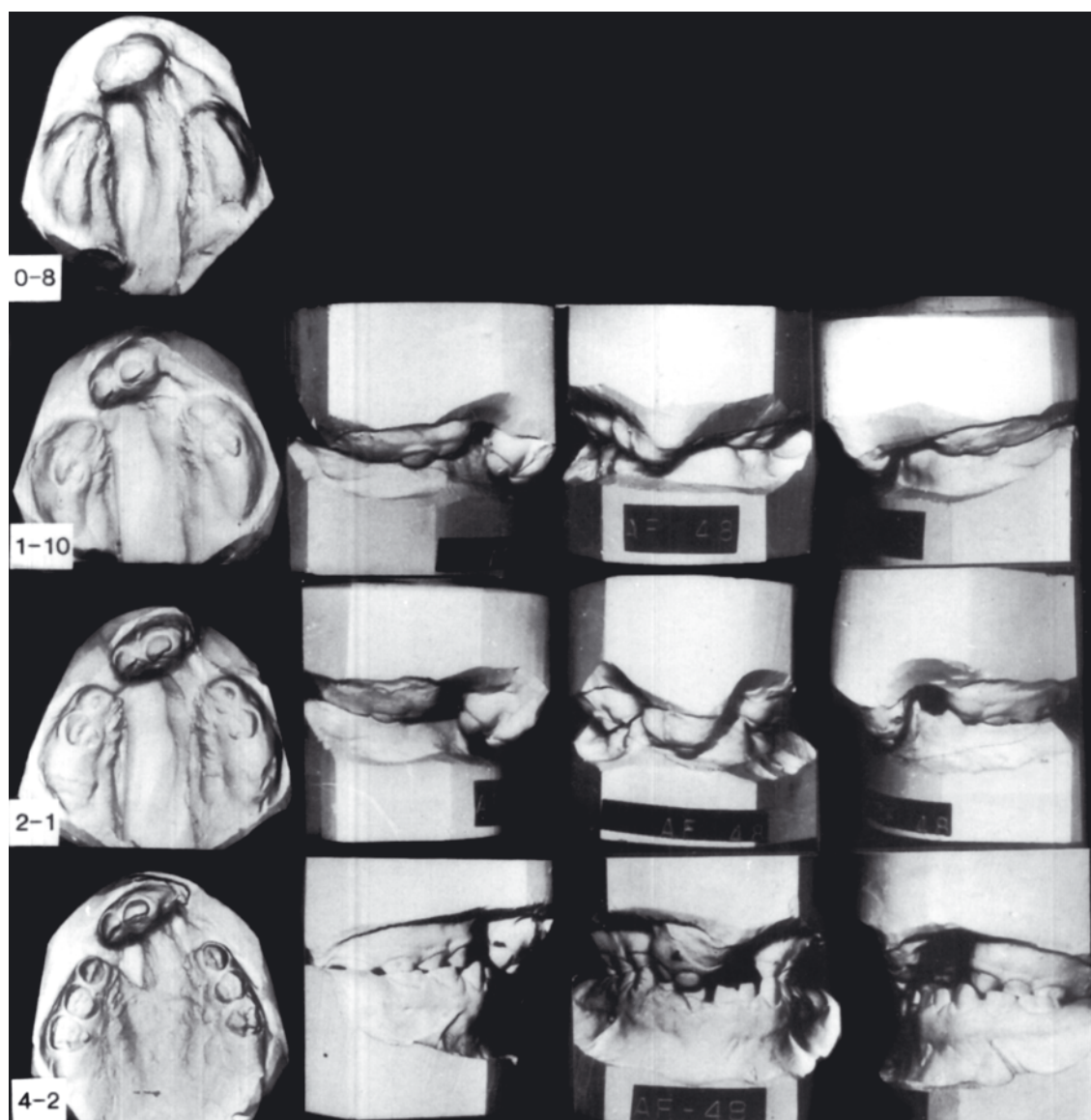


Fig. 6C.44. Case CS (AF-48). Serial casts show that the extreme premaxillary protrusion with large anterior cleft space at birth is still present at 6-1 years of age

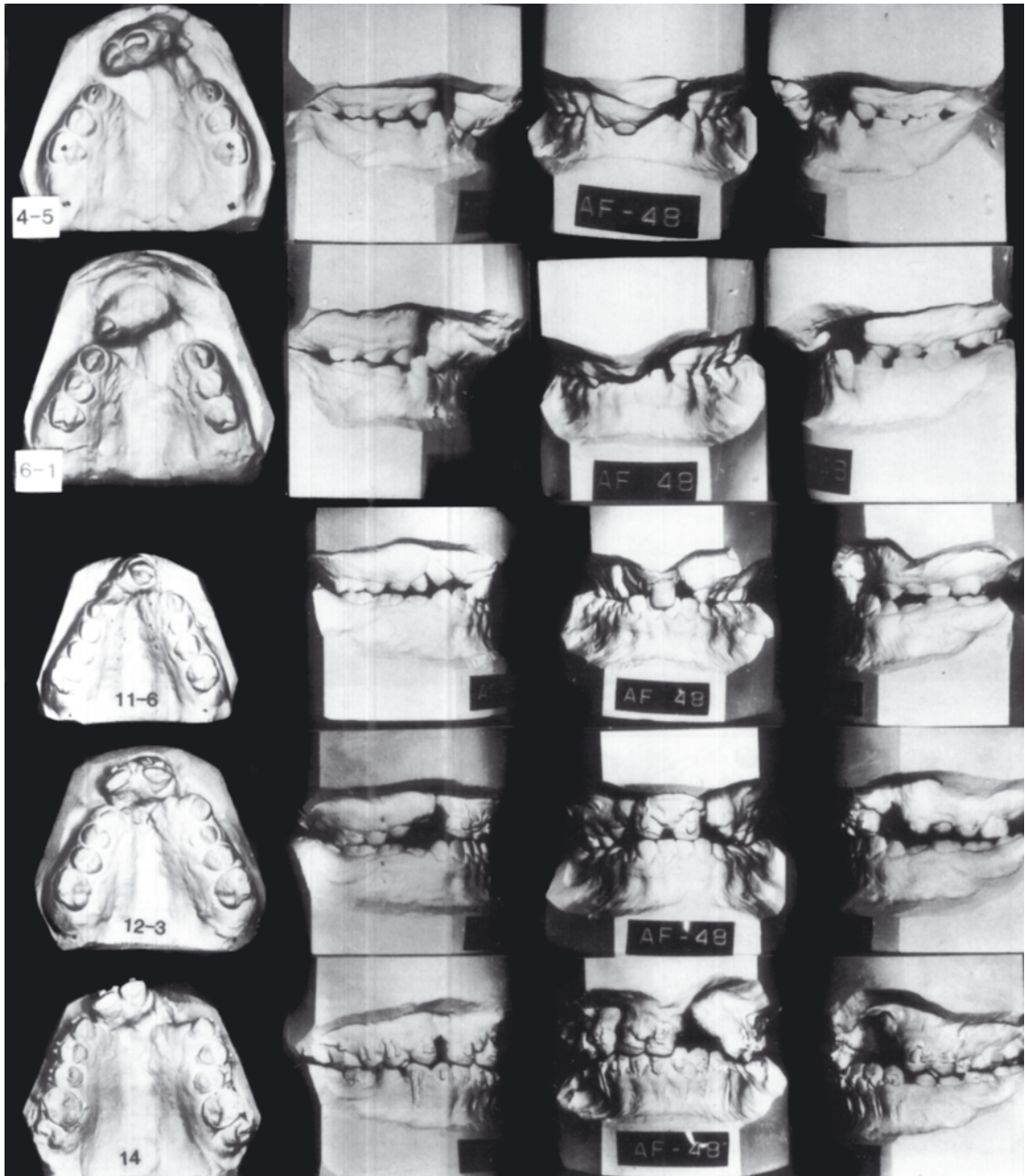


Fig. 6C.44. (continued) 11-6 With increased facial growth, the increasing tonicity of the buccal muscle forces collapsed the maxillary arch placing the posterior teeth in crossbite. The premaxilla is now upright and in an acceptable overbite-overjet relationship. 12-3 Maxillary expansion has been initiated. The

maxillary central incisors are in tip-to-tip relationship. 14 Continued orthodontic treatment to advance the premaxilla and position the incisor teeth in proper overjet-overbite relationship

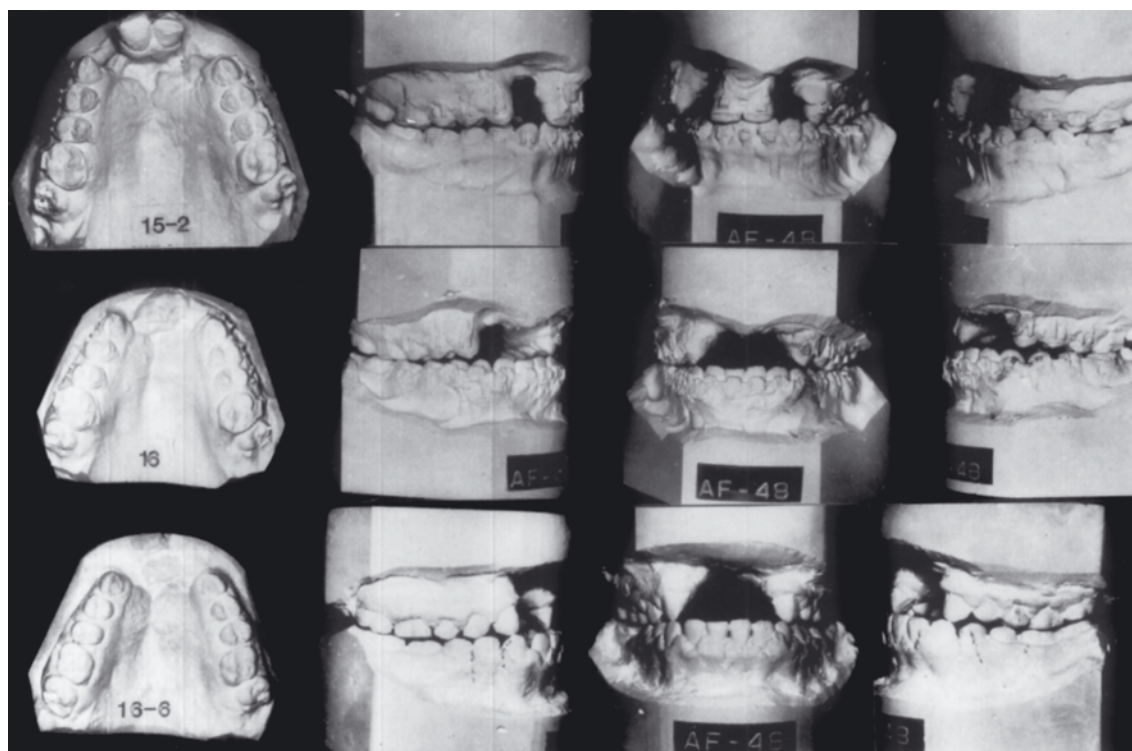


Fig. 6C.44. (continued) 15-2, 16, and 16-6 Premaxillary repositioning with soft tissue closure of the anterior cleft space was unsuccessful. As a result of the premaxillary central incisors showing external root absorption and loss of periodontal support, they were extracted and the oronasal opening closed with

adjacent soft tissue. A removable maxillary prosthesis replaces the missing teeth and bumpers the upper lip. Comment: As already suggested, the treatment of choice is to reposition the lateral palatal segments anteriorly while leaving the premaxilla as is

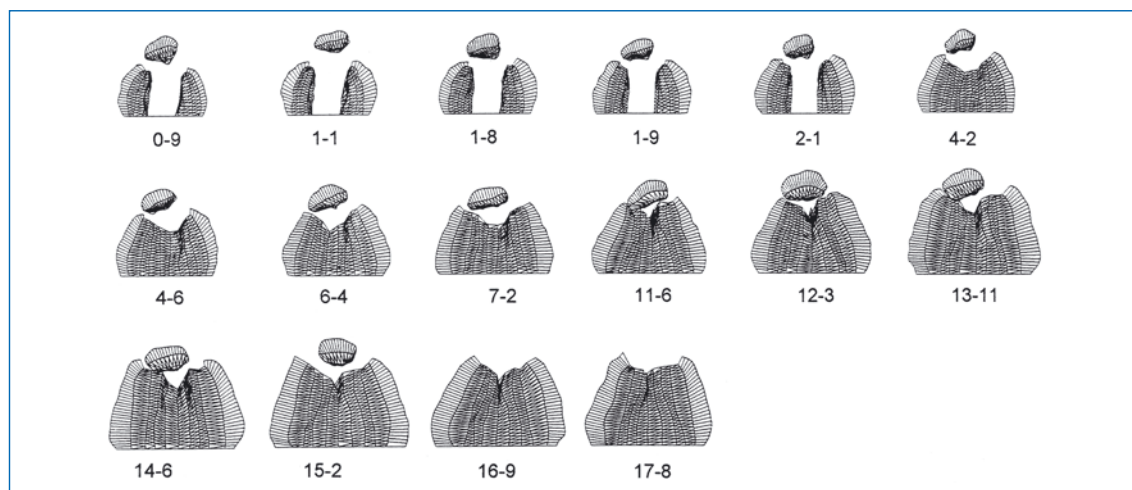


Fig. 6C.45. Case CS (AF-48). Computerized tracings of serial casts drawn to scale. This shows the lack of palatal growth and reduction in cleft space over 2 years prior to surgical closure of the palatal cleft. The anterior cleft space remains large up to 15-2 years. 16-9 and 17-8 The premaxillary incisors were extracted. Comment: This case clearly demonstrates the severe

degree of osteogenic deficiency that can exist in bilateral clefts of the lip and palate. The once protruding premaxilla can become retrusive with growth (time) and may eventually need to be brought forward. Although palatal growth does occur, it may not be sufficient to appreciably reduce the posterior cleft space

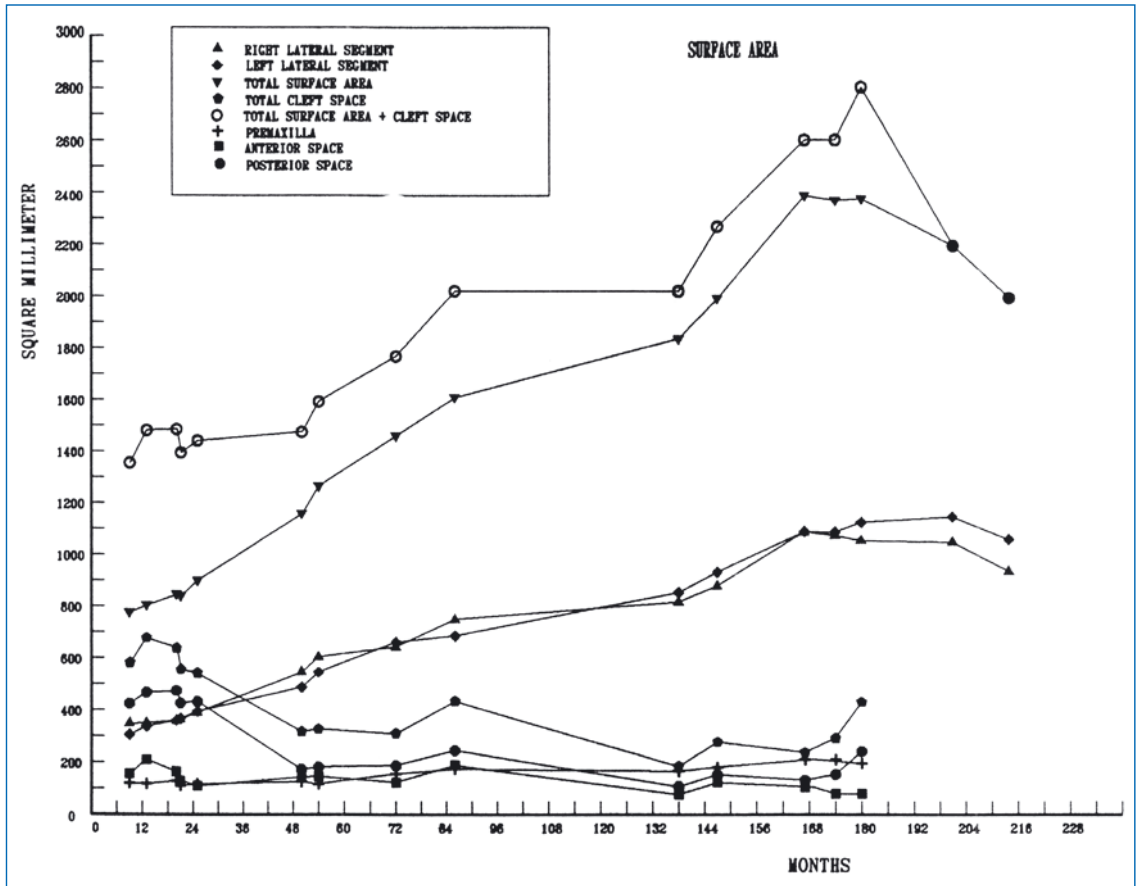


Fig. 6C.46. Case CS (AF-48). The palatal segments show a very gradual growth acceleration curve, while the posterior cleft space gradually reduces in size

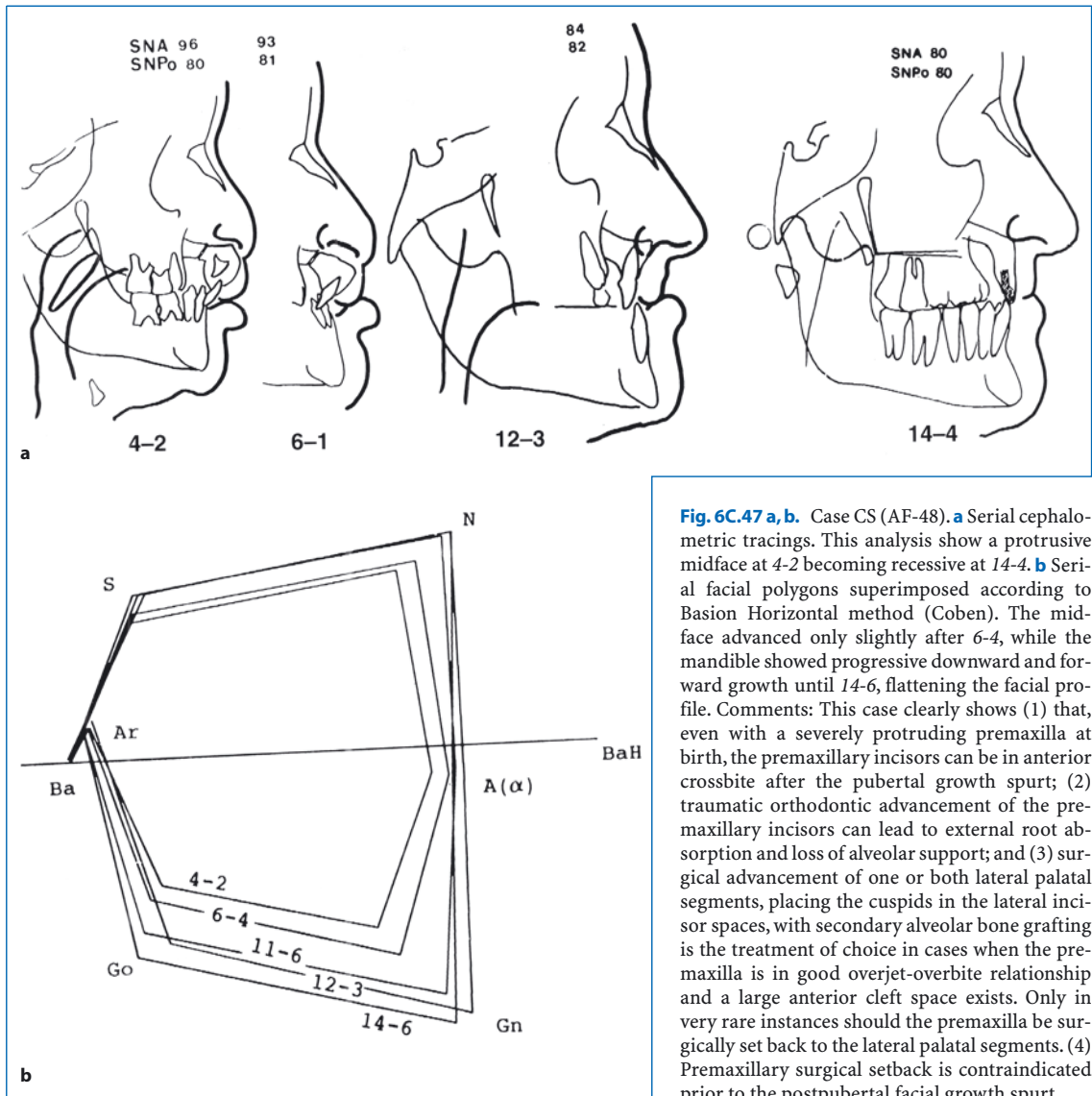


Fig. 6C.47 a,b. Case CS (AF-48). **a** Serial cephalometric tracings. This analysis shows a protrusive midface at 4-2 becoming recessive at 14-4. **b** Serial facial polygons superimposed according to Basion Horizontal method (Coben). The midface advanced only slightly after 6-4, while the mandible showed progressive downward and forward growth until 14-6, flattening the facial profile. Comments: This case clearly shows (1) that, even with a severely protruding premaxilla at birth, the premaxillary incisors can be in anterior crossbite after the pubertal growth spurt; (2) traumatic orthodontic advancement of the premaxillary incisors can lead to external root absorption and loss of alveolar support; and (3) surgical advancement of one or both lateral palatal segments, placing the cuspids in the lateral incisor spaces, with secondary alveolar bone grafting is the treatment of choice in cases when the premaxilla is in good overjet-overbite relationship and a large anterior cleft space exists. Only in very rare instances should the premaxilla be surgically set back to the lateral palatal segments. (4) Premaxillary surgical setback is contraindicated prior to the postpubertal facial growth spurt.

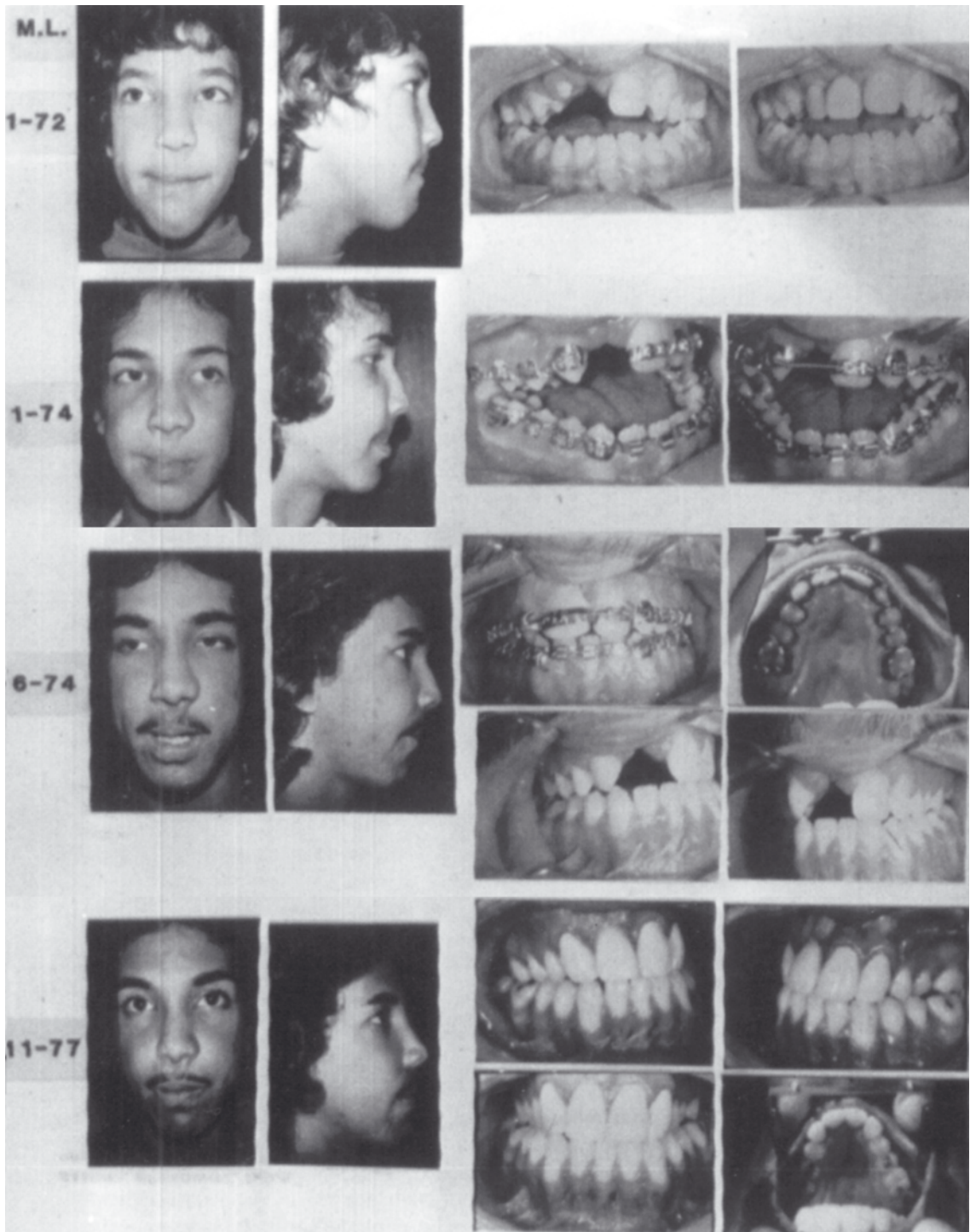


Fig. 6C.48. Case ML demonstrates poor facial growth pattern in a BCLP leading to a retruded midface with a severe anterior open bite. This patient came to the clinic at 5 years, 7 months of age with an anterior open bite. The upper lip was long and tight with the lateral elements brought together below the prolabium. The palatal cleft was closed at 12 months of age. At 13-10, a removable plate replaced the right central incisor. At 15-11,

orthodontic preparation for Lefort I posterior-impaction. At 16-4, after maxillary surgery. At 19-9, after anterior fixed bridge used to stabilize the palatal segments and replace missing teeth. The changes in occlusion and total facial height led to a more relaxed soft tissue profile. With reduction in lower vertical facial height, the upper to lower lip position became more aesthetic.

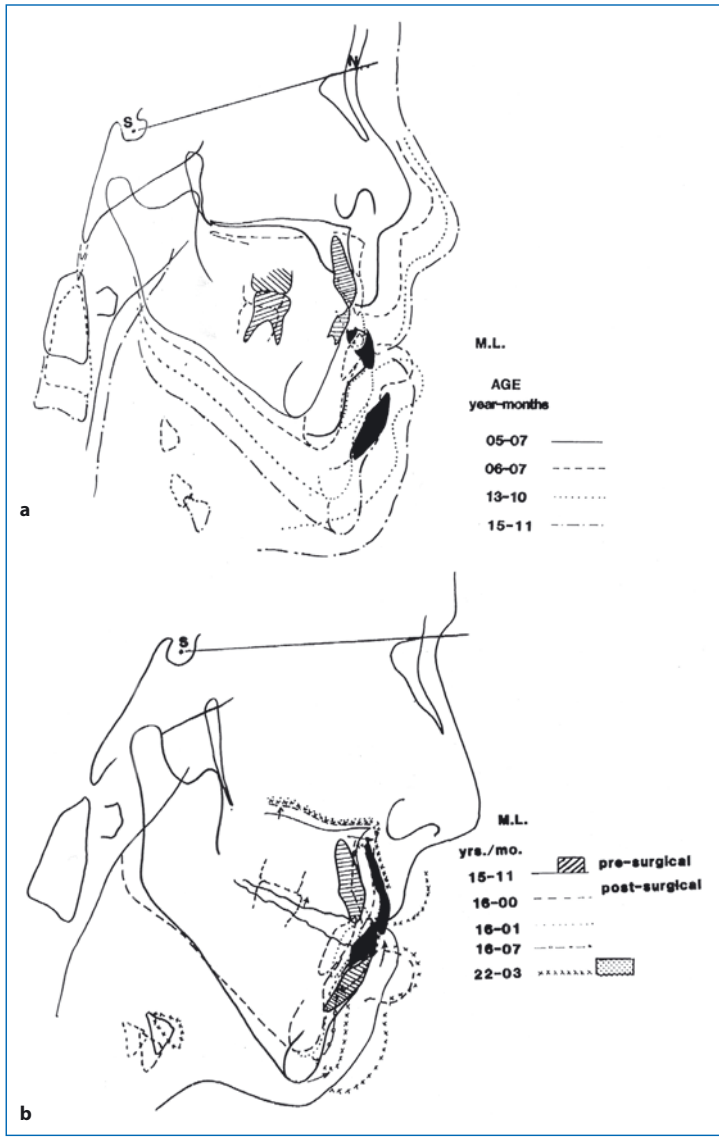


Fig. 6C.49 a, b. Case ML. Superimposed serial cephalometric tracings on SN and registered at S before surgery. **a** Between 5-7 and 15-11 the face demonstrated a very poor growth pattern associated with diminished midfacial growth. The incisors were in tip-to-tip relationship at 5-7; by 6-7 the anterior open bite with a retruded midface was present, which became worse with time. **b** Serial cephalometric tracing superimposed on SN and registered at S, after posterior impaction and maxillary advancement. The mandible autorotated closing the anterior open bite. The curvature to both lips became more prominent and aesthetic. Comment: Placing the lateral lip elements below the prolabium creates a long and tight upper lip. Maxillary growth disturbance is always expressed in all three dimensions, resulting in diminished vertical and horizontal growth increments. Maxillary advancement with slight posterior impaction causes mandibular autorotation, bringing the lower incisors upward and anteriorly closing the anterior open bite. The middle and lower vertical facial heights became more harmonious

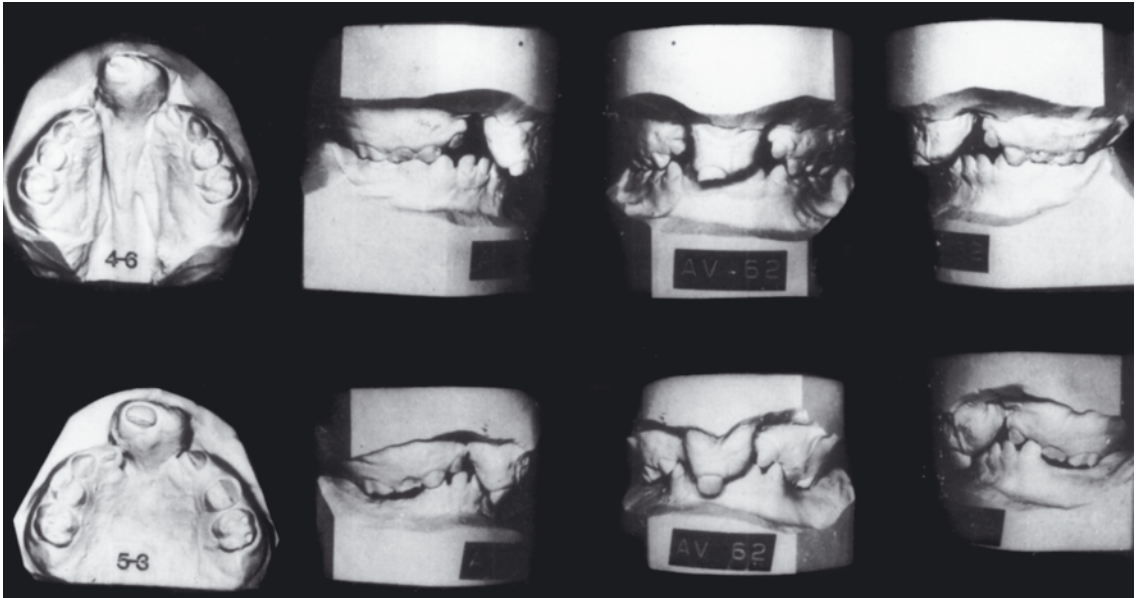


Fig. 6C.50. Case CT (AV-62) demonstrates a severe premaxillary overjet leading to a retruded midface and the successful use of protraction orthopedics. This patient was adopted from Central American parents. 4-6 It is important to note the wide cleft

space with an excellent Class I buccal occlusion prior to surgical cleft closure. 5-3 Two months after palatal cleft closure, the occlusion is still acceptable

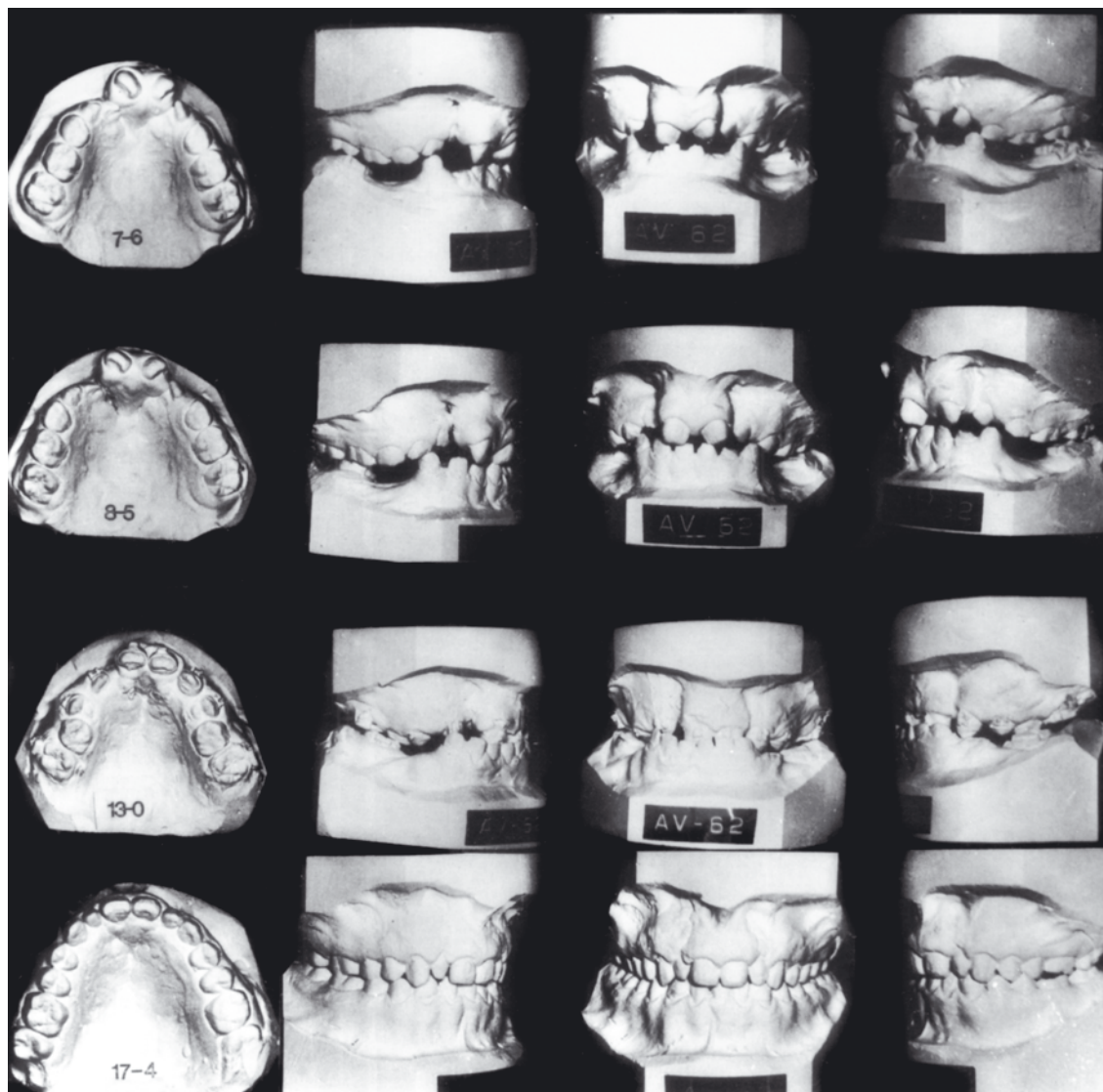


Fig. 6C.50. (continued) 7-6 and 8-5 A secondary alveolar bone graft was placed at 8-0. The anterior teeth are in a tip-to-tip relationship with the buccal occlusion in crossbite. 13-0 Complete anterior and buccal crossbite. Preparation is made to begin orthodontic treatment to expand the maxillary arch as well as to advance the maxillary complex with protraction mechanics. 17-4 After orthodontic treatment. A lower central incisor was extracted to allow for incisor, retraction. Excellent occlusion is retained with a Hawley appliance. Comment: This case clearly demonstrates that, in complete clefts of the lip and palate, the lateral palatal segments at birth can be in good relationship with the lower arch and not collapsed, as McNeil suggests. In

most cases, the surface area of the lateral palatal segments can be considered to be too small when compared to the cleft size. Too early cleft closure using a von Langenbeck procedure will leave large areas of uncovered bone when the mucoperiosteum is moved medially. Denuded bone undergoes epithelialization with scarring, which in turn interferes with palatal growth in all three dimensions, leading to the buccal and anterior crossbite. In this case, protraction mechanics were successful. However, because the ideal overjet-overbite relationship could not be achieved, it was necessary to remove a lower central incisor to retract the anterior segment. This benefited the buccal occlusion as well.

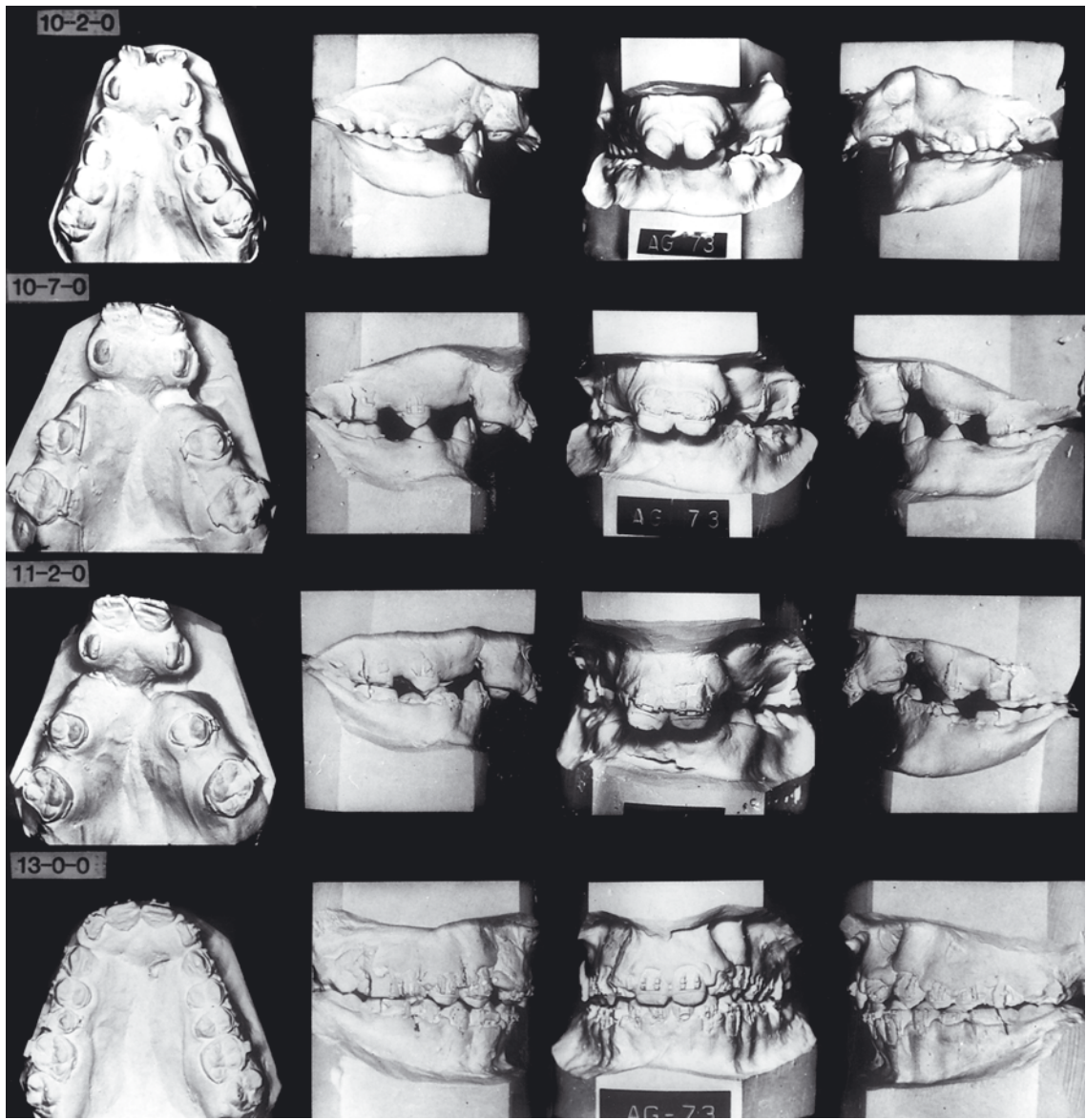


Fig. 6C.51. Case RB (AG-73). Serial dental casts. Closure of a large anterior cleft space in a BCLP by bringing impacted cuspids into position and aligning the lateral incisors. 10-2 Class II malocclusion in the mixed dentition with a severe overjet and overbite. Bilateral deciduous cuspid crossbite with no anterior cleft space. 10-7 With loss of the deciduous cuspid and its supporting alveolar bone along with the horizontal impaction of the permanent cuspids, there was deficient alveolar bone between the lateral palatal segments and the premaxilla, creating

a large anterior cleft space. 11-2-0 Waiting for the eruption of permanent bicuspid before initiating orthodontics. 13-0-0 After the horizontally impacted cuspids and their supporting alveolar bone and the lateral incisors were brought within the arch, the premaxillary overjet was reduced. Both of these factors were responsible for the closure of the anterior cleft space. This was followed by a secondary alveolar cranial bone graft to close fistulae and remaining cleft spaces

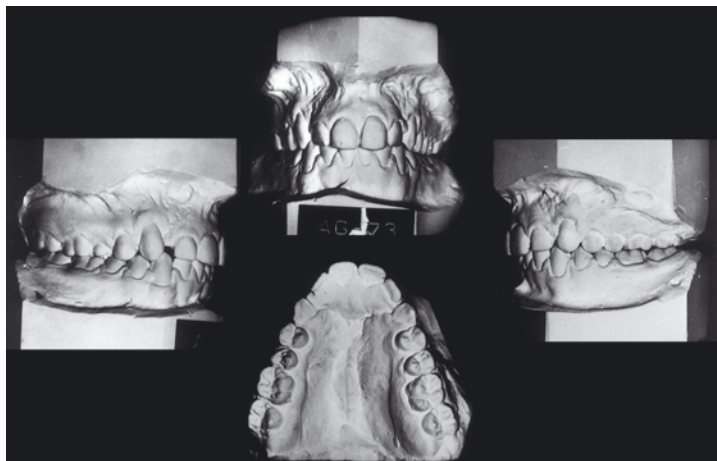


Fig. 6C.51. (continued) 13-2 Final occlusion with a full complement of teeth. Comment: The clinician must always consider the importance of the alveolar bone supporting the teeth whether they are in alignment or malpositioned. When all of the teeth in the lateral palatal segments can be accommodated within the arch, the anterior cleft space can usually be closed without an osteotomy

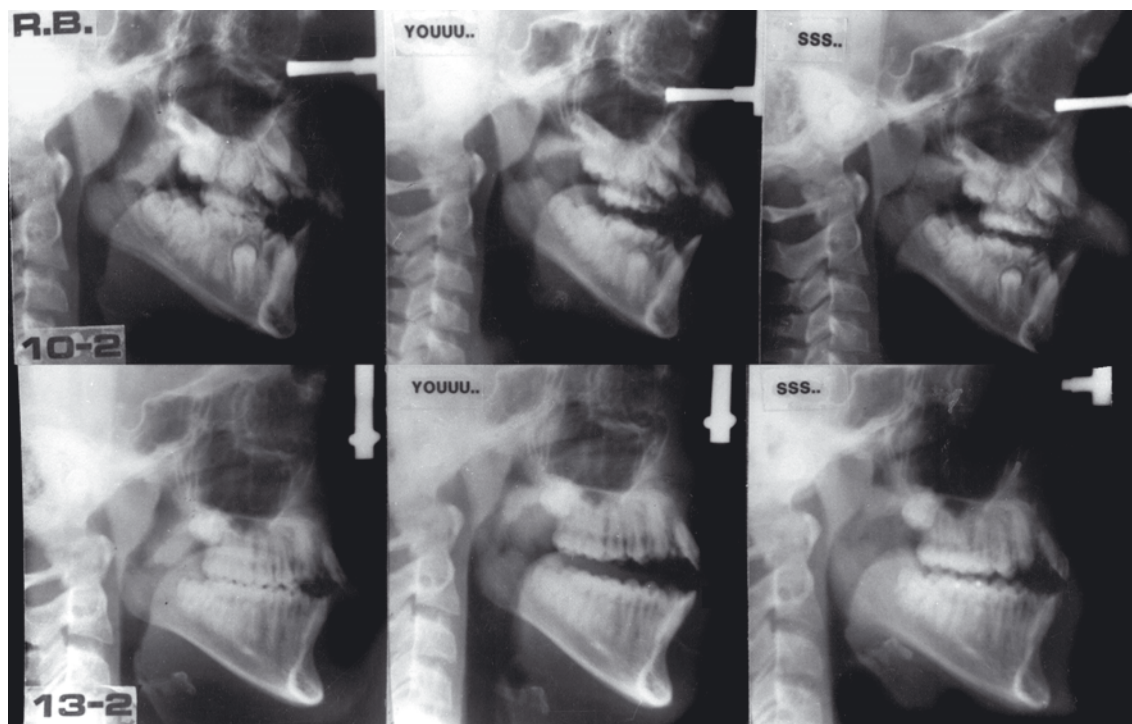


Fig. 6C.52. Case RB. Cephalometric view of the pharyngeal at rest and with space and velar elevation. 10-2 Left: At rest, and when vocalizing “Youu . . .” Although there is good velar elevation, the velum does not make contact with the adenoid. Right: Vocalizing “Sss . . .” This sound produces velar stretch, permitting contact to be made with the adenoid. 13-2 Good velar ele-

vation when vocalizing “Youu . . .” and “Sss . . .”. Comment: Cephalometric evaluation is necessary to view the structure of the pharyngeal space (see Chaps. 11 and 12). This is not a functional test to evaluate velopharyngeal competency, but it may give some insight into why velopharyngeal incompetency may exist



Fig. 6C.53 a-r. Case ES (EE-34) demonstrates excellent facial growth pattern in a IBCLP showing good midfacial growth. An external elastic off a head bonnet was used to ventroflex the premaxilla prior to lip surgery. The lip was united at 5 months of age using a Fork flap procedure. The palatal cleft was closed at 43 months of age using a modified von Langenbeck procedure. **a** and **b** Face and palate at birth. **c** and **d** 3 months after lip surgery. **e-i** 5 years of age with excellent buccal occlusion and good overbite-overjet relationship of the anterior teeth. Upper lip is protrusive



Fig. 6C.53 a-r. (continued) **j** and **k** 10 years of age after secondary alveolar bone grafting. **l** and **m** A left cuspid crossbite resulted from secondary alveolar bone graft surgery



Fig. 6C.53 a-r. (continued) **n-r** 16 years. After orthodontics was completed. The left lateral incisor erupted through the bone graft; the right lateral incisor is missing. The arch form is main-

tained with a removable Hawley retainer, which carries a lateral incisor replacement

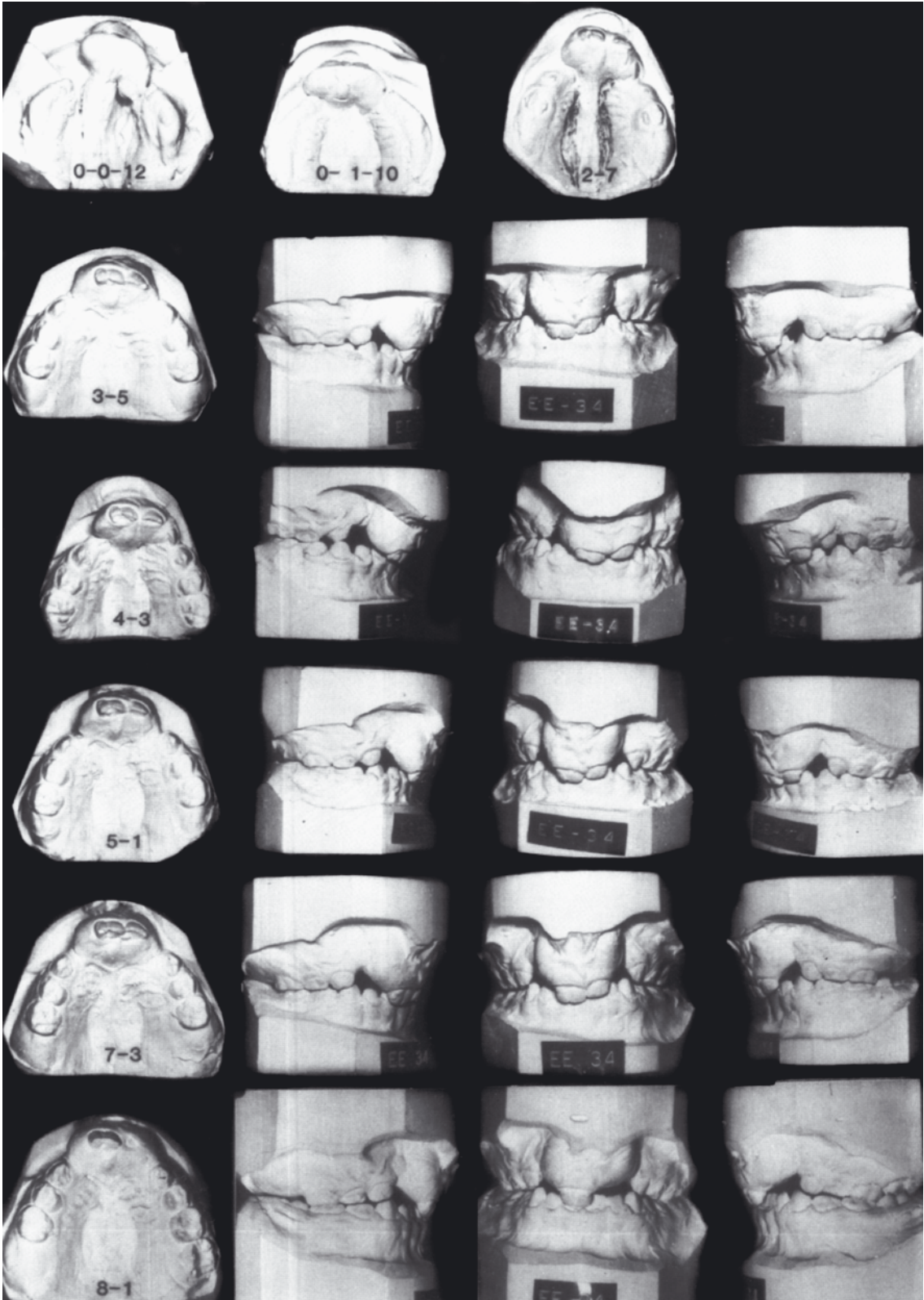


Fig. 6C.54.

Fig. 6C.54. Case ES (EE-34). Serial dental casts. 0-0-12 At birth. 0-1-0 After uniting the lip, the premaxilla has been positioned adjacent to the lateral palatal segments. 2-7 The palatal cleft is still open. Tongue force has moved the premaxilla forward. 3-5 After palatal cleft closure there is excellent buccal occlusion. 4-3, 5-1, 7-3, and 8-1 Good buccal and anterior occlusion is present for the next 4 years

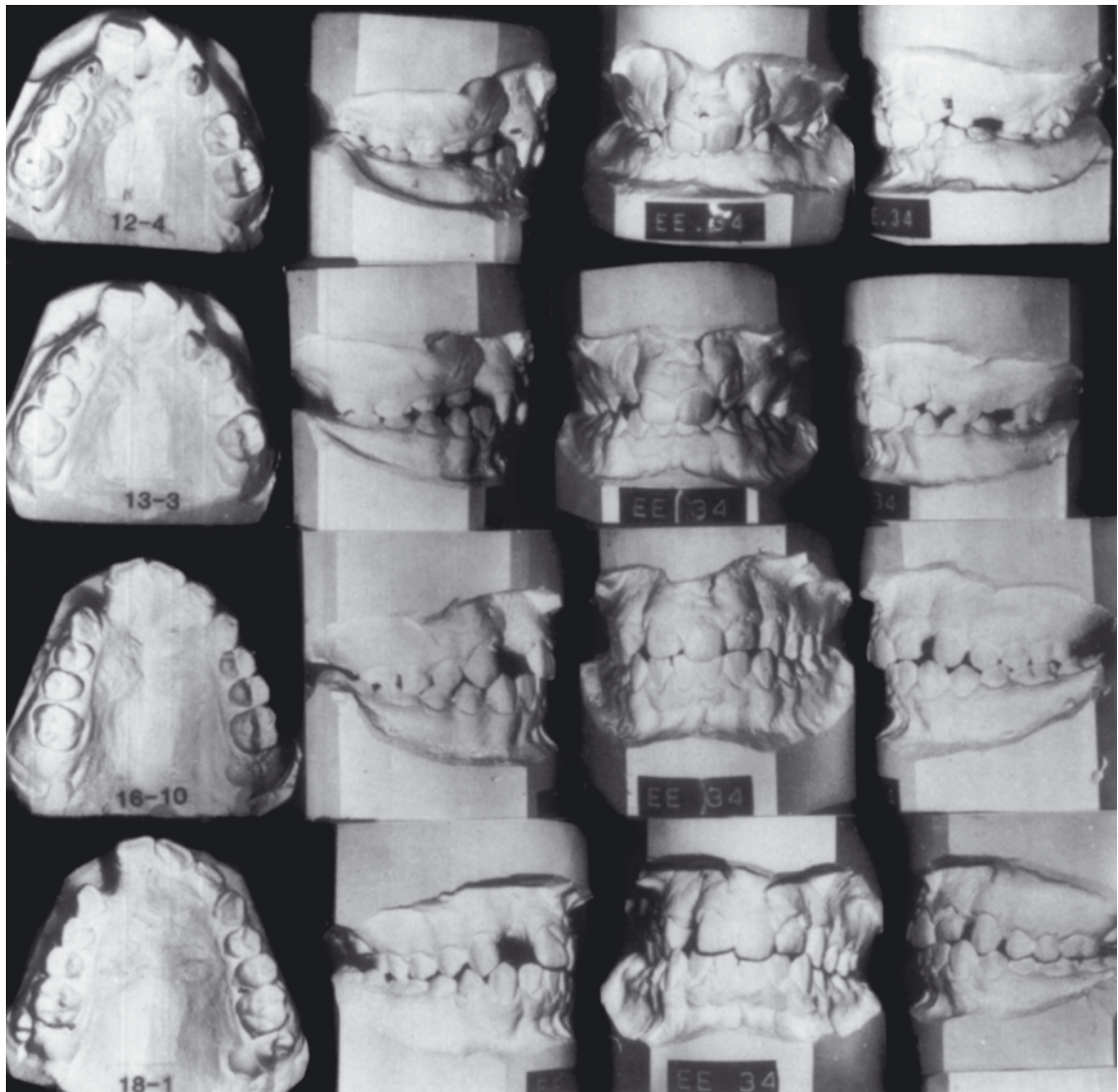


Fig. 6C.54. (continued) 12-4 and 13-3 This crossbite remains for the next 3 years and does not hinder dental function. 16-10 and 18-1 After orthodontics. Good Class I occlusion with good anterior overjet and overbite

EE - 34

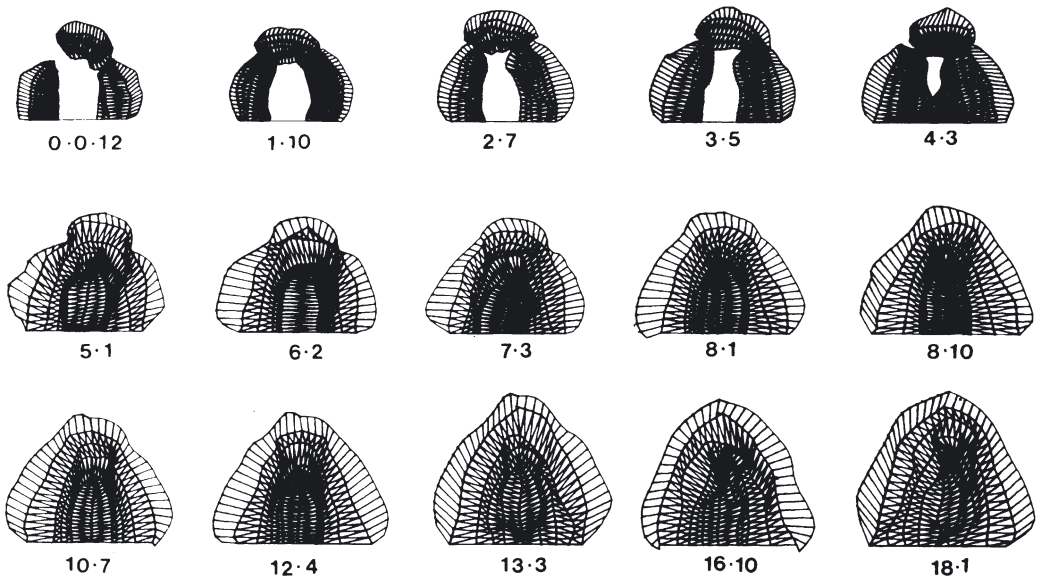


Fig. 6C.55. Case ES (EE-34). Computer-generated images of serial palatal casts. All of the casts are in proportion. Palatal growth occurs on all surfaces but mainly transversely and posteriorly to accommodate the unerupted molars. Although the palatal surface is gradually increasing in size, the posterior cleft

space remains almost constant in size over the same period of time. When the palatal cleft was closed at 43 months, more mucoperiosteal tissue was available to prevent growth-inhibiting scar tissue from being created

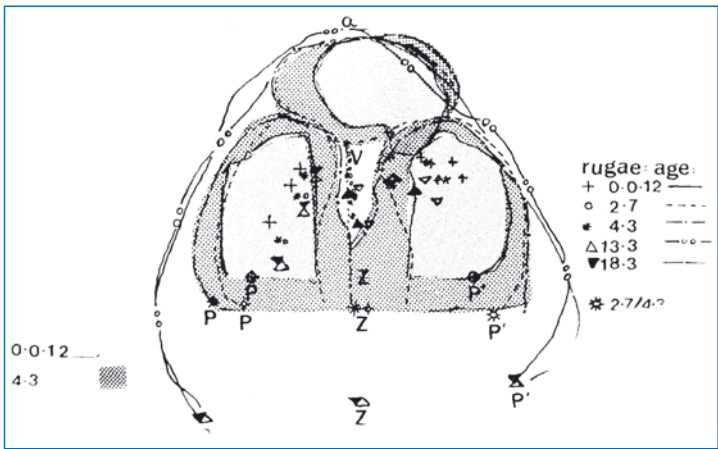
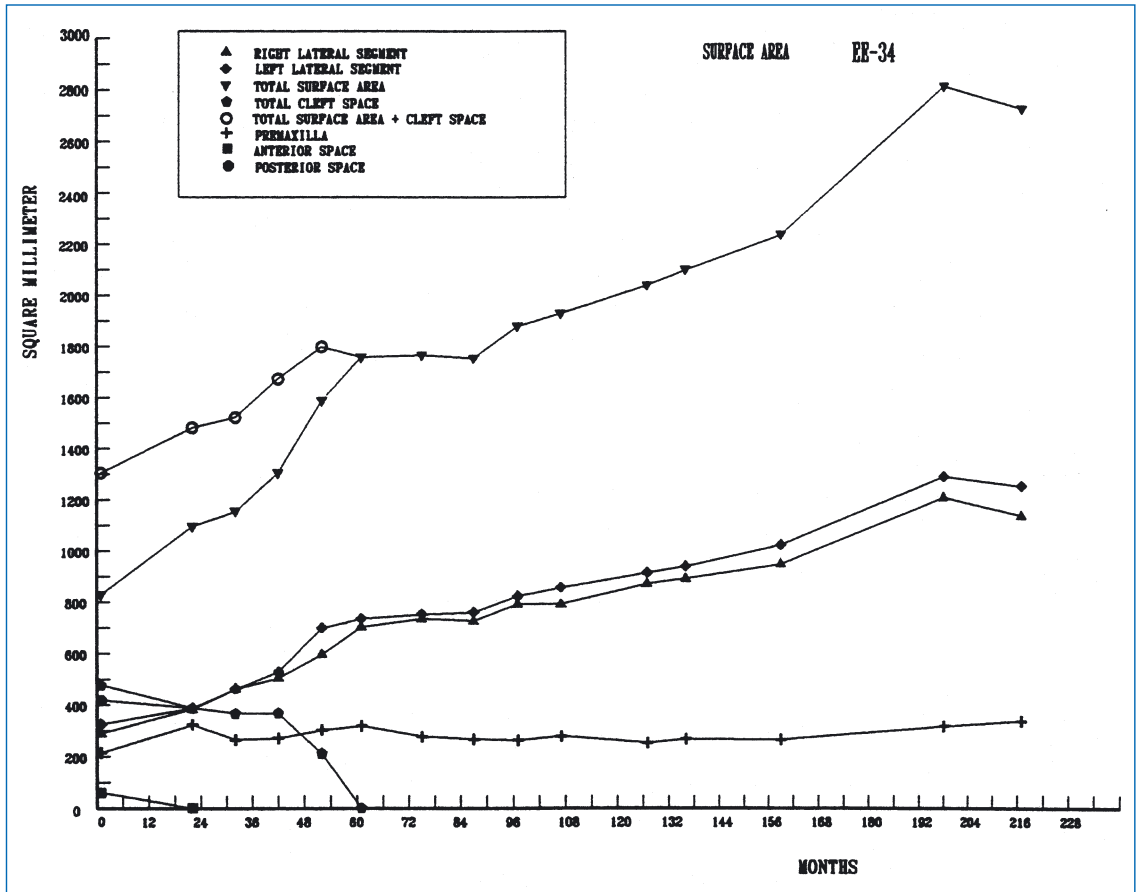


Fig. 6C.56. Case ES (EE-34). Computer-generated outlines of palatal casts superimposed on the palatal rugae. This study demonstrates that palatal growth occurs mainly in the posterior and transverse areas with very little premaxillary growth. More significantly, it shows that the premaxilla is relatively stable in its geometric position within the maxillary complex, that is, the face grows up to and around the original position at birth



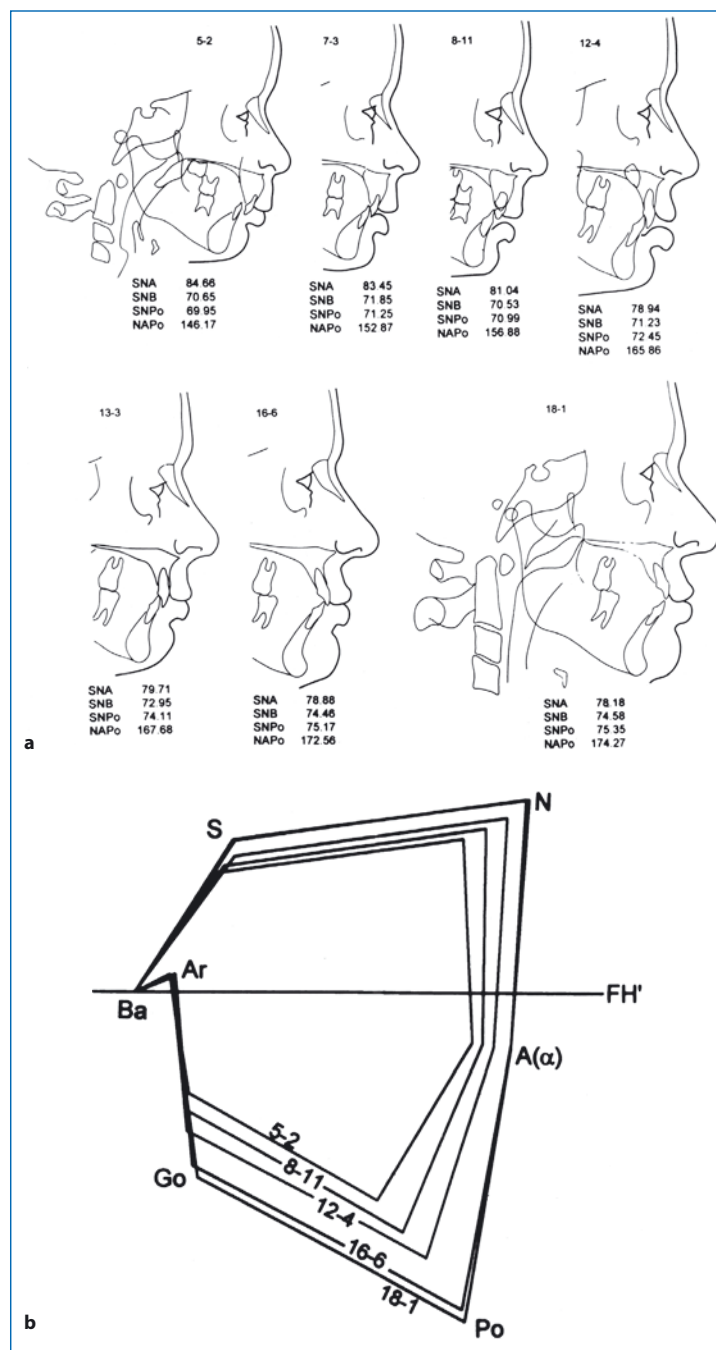


Fig. 6C.58 a,b. Case ES (EE-34). **a** Serial cephalometric tracings show ideal facial measurements evolving. A superior-based pharyngeal flap is outlined in 18-1. **b** Serial polygons superimposed according to the Basin Horizontal method (Coben). This series demonstrates an ideal facial growth pattern and reflects the use of physiological maxillary surgery: (1) The anterior cranial base shows good growth increments. (2) Midfacial growth increments are continuous and only slightly less than those seen at *N* (Nasion). (3) Mandibular growth shows more forward than downward changes which is conducive to the flattening of the facial profile. Comment: Taken together, all of these growth changes are conducive to the flattening of the facial profile. Such extensive midfacial growth is usually not seen in BCLP cases with such a large cleft space relative to the palate's surface area that have had the palatal cleft closed earlier than 43 months of age. We speculate that this finding strongly suggests that: (1) delaying palatal surgery to 3 years of age may be more conducive to good midfacial growth in some cases than early (before 1 year) palatal closure. However, we believe that it is not necessary to postpone palatal closure to after 5 years of age in all cases. (2) Hypotonic lip pressure has not exerted sufficient pressure through the premaxilla to the premaxillary vomerine suture to reduce its growth before 16-6. However, between 16-6 and 18-1 midfacial growth has ceased

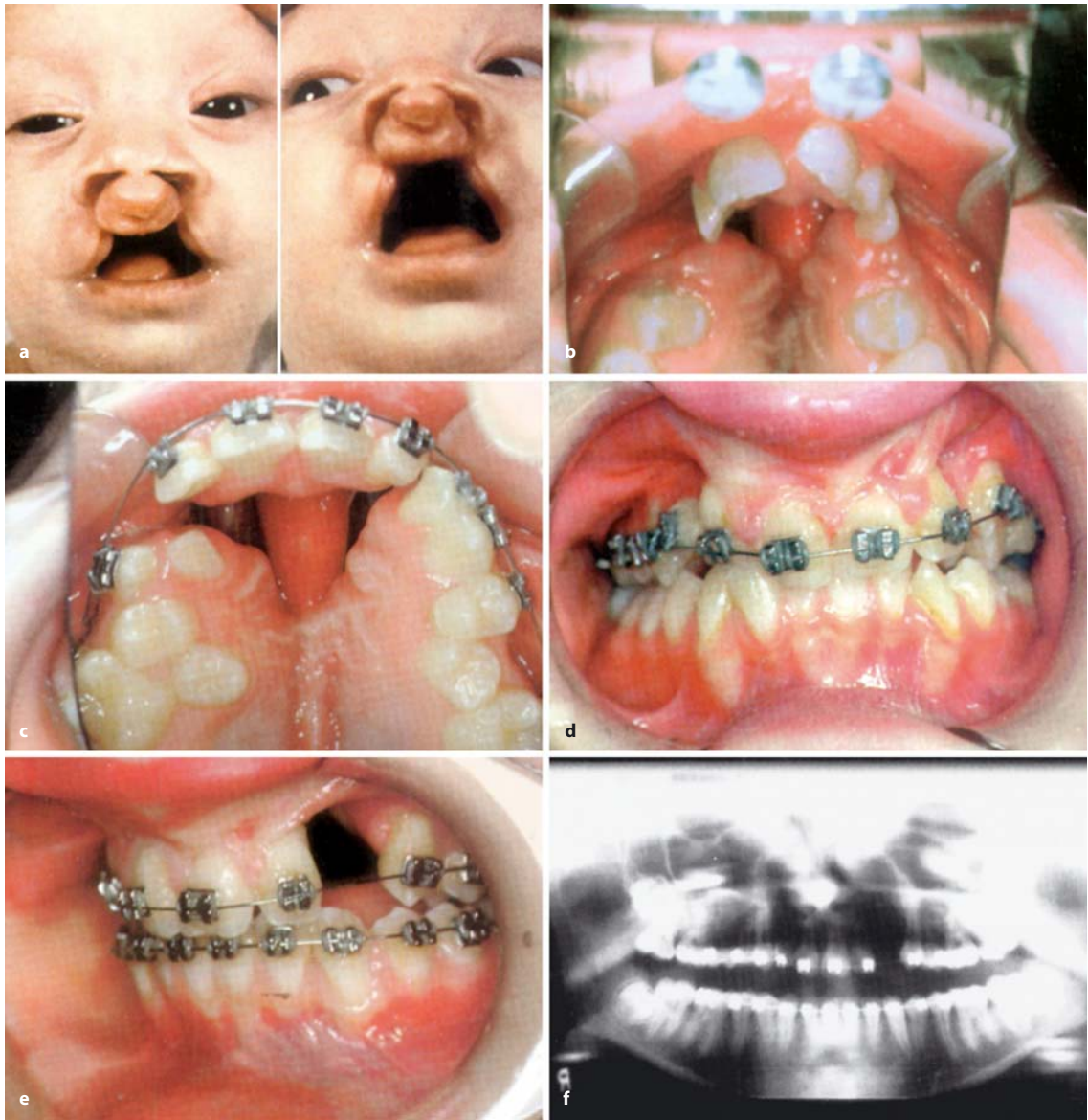


Fig. 6C.59 a–n. Anterior positioning of the lateral palatal segments in an adolescent with CBCLP to close a large anterior cleft space (Courtesy of [72]) A 16-year-old with repaired BCLP underwent combined orthognathic-orthodontic procedure to align the teeth, close the large cleft space while maintaining the existing premaxillary overjet-overbite relationship. **a** Initial cleft lip and palate deformity at 8 weeks of age. **b** Palatal view at

10. **c** and **d** Preoperative palatal and frontal view at 15 years showing large anterior cleft space and dental crowding of the lateral palatal segments. The right second bicuspid is to be extracted. **e** Preoperative occlusal view showing large cleft space with posterior positioned lateral palatal segment. **f** Preoperative panorex at 17 years of age

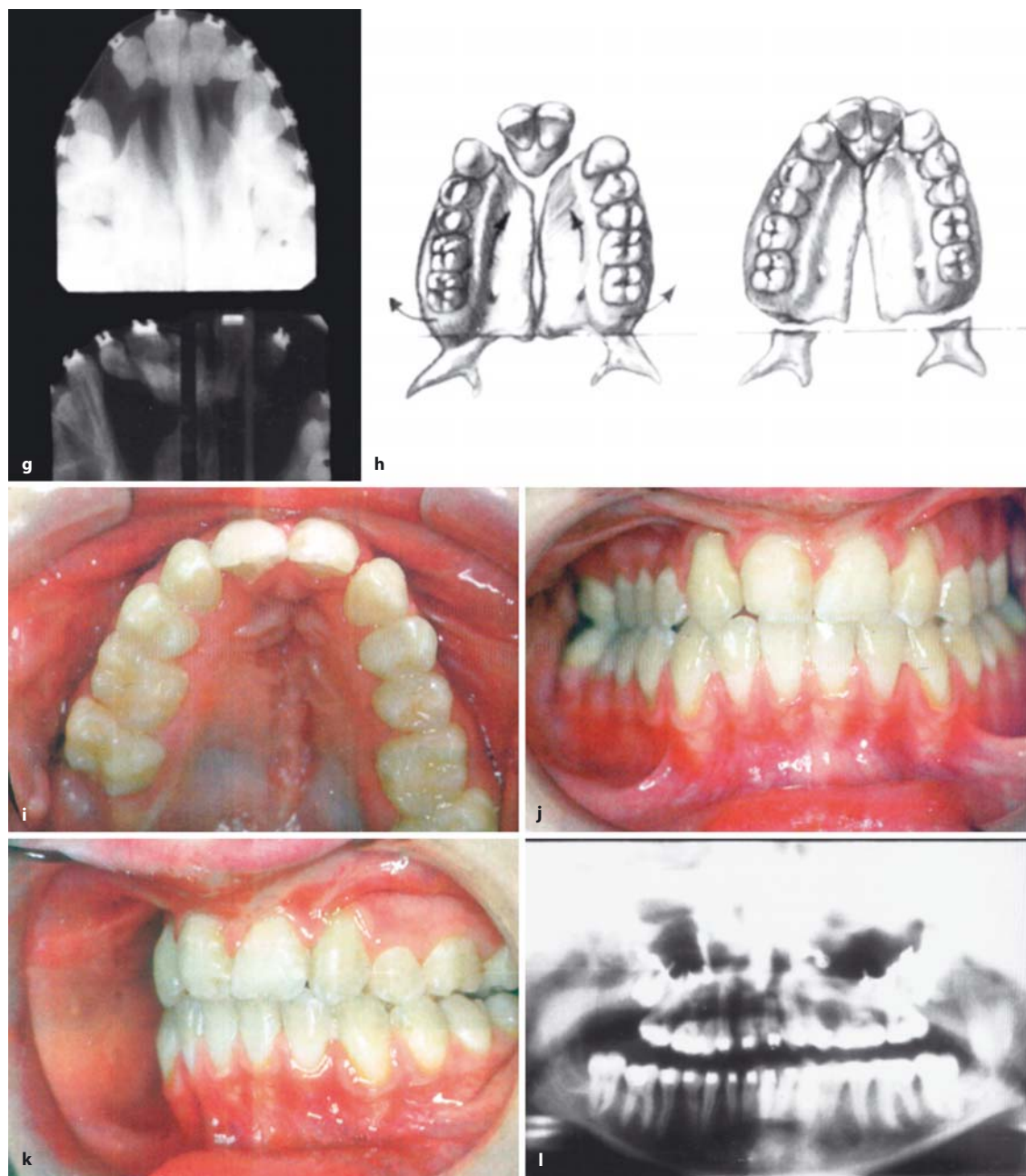


Fig. 6C.59 a-n. (continued) **g** Occlusal radiograph. **h** Line drawing of the proposed palatal surgery showing the posterior widening and anterior movement of the lateral palatal seg-

ments. **i**, **j**, and **k** Postoperative intraoral photographs. **l** Postoperative panorex

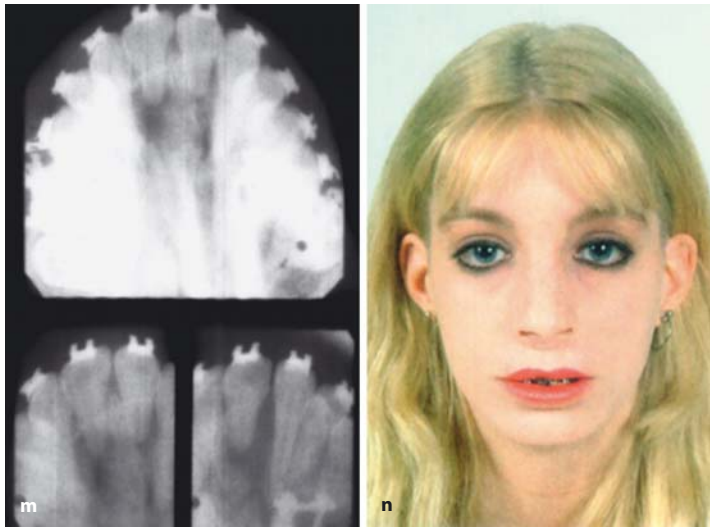


Fig. 6C.59 a–n. (continued) **m** Occlusal radiographs. **n** Postoperative frontal facial view. Comment: There was limited alveolar bone support to the lateral incisor areas. The premaxilla was mobile. Marked velopharyngeal incompetence with regurgitation of fluid while drinking and intake of air, while speaking required a palatal obturator. The surgical plan was to extract the questionable lateral incisors and reposition the maxillary lateral segments to close the lateral incisor and large anterior cleft spaces

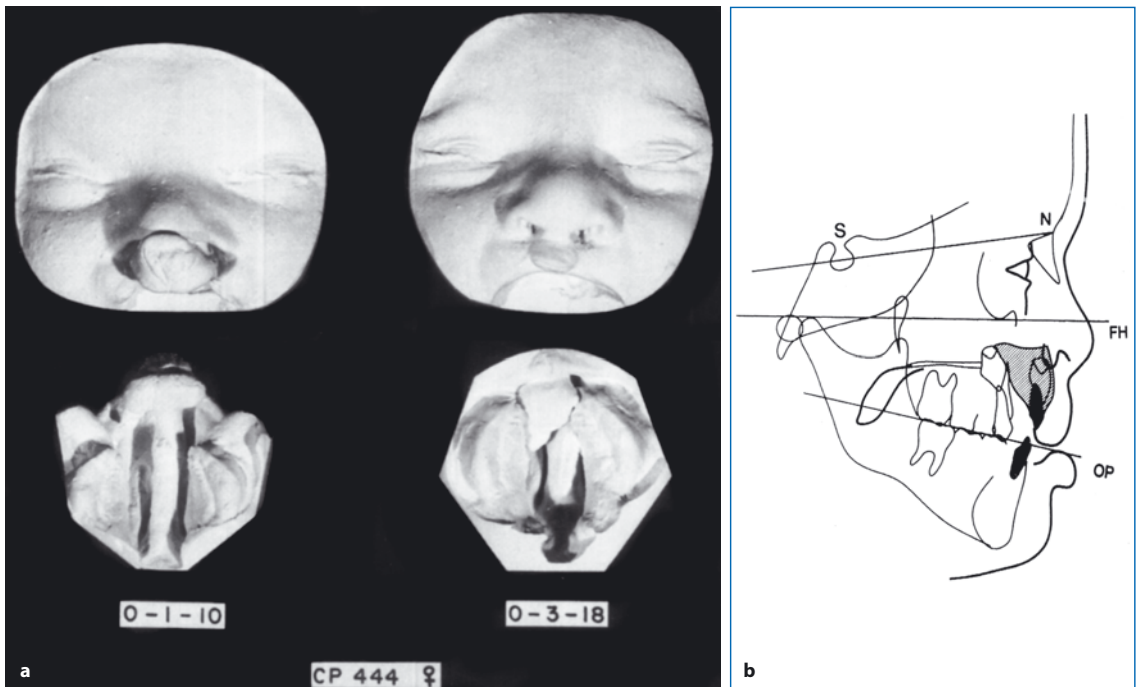


Fig. 6C.60 a, b. Case CP (444) demonstrates the effects of early premaxillary surgical setback. The purpose of this surgical procedure is to establish ideal arch form prior to lip surgery. The procedure lost favor when surgeons found that it led to severe midfacial growth retardation and an anterior open bite. **a** Before and after premaxillary setback. The premaxilla is posi-

tioned within the arch. **b** 4 years later, the premaxilla, having been detached from the vomer, fails to descend with the palate with growth, creating an anterior open bite. Orthodontics in the permanent dentition will super erupt the maxillary incisors with only slight change in maxillary position (Courtesy of S. Pruzansky)

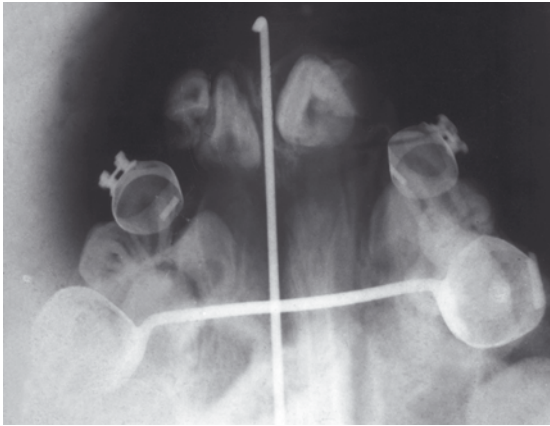


Fig. 6C.61. A Kirschner wire that penetrated dental sacs of the central incisors, causing tooth malformation. Due to the spatial relationship of the premaxilla to the vomer, the Kirschner wire frequently fails to enter the vomer as seen here

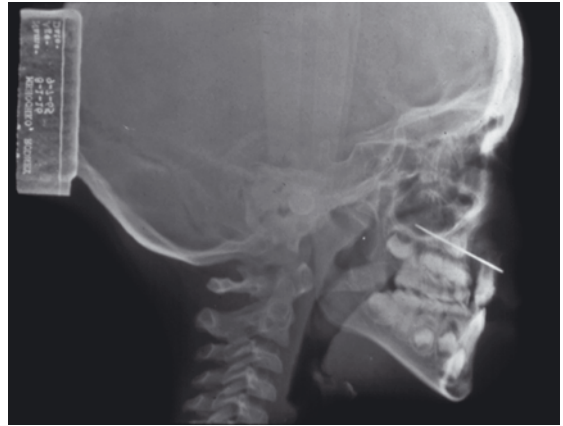


Fig. 6C.62. Lateral cephalograph showing a malpositioned Kirschner wire. After the premaxilla is surgically retropositioned, surgeons used to attempt to stabilize it by placing wires of various sizes through the premaxilla into the vomer. The procedure has not been successful and should be abandoned because it can devitalize the teeth in the premaxilla and the wire can migrate to various areas within the skull

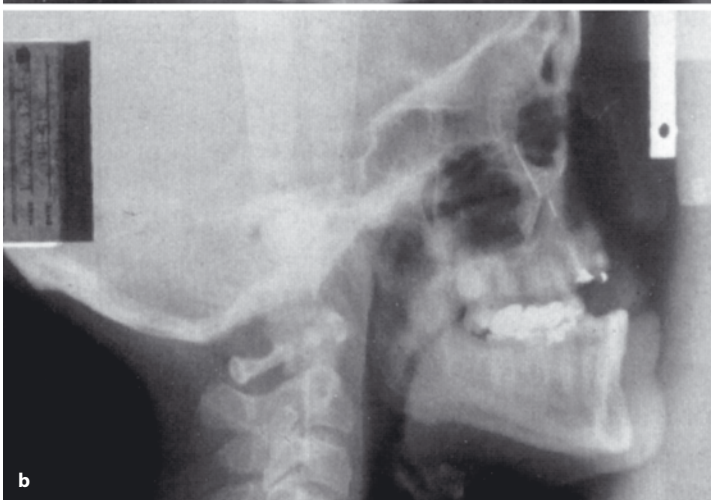


Fig. 6C.63 a, b. Two views of a Kirschner wire 35 years after insertion. **a** Panorex and **b** lateral cephalometric films show the displaced Kirschner wire located high in the vomer

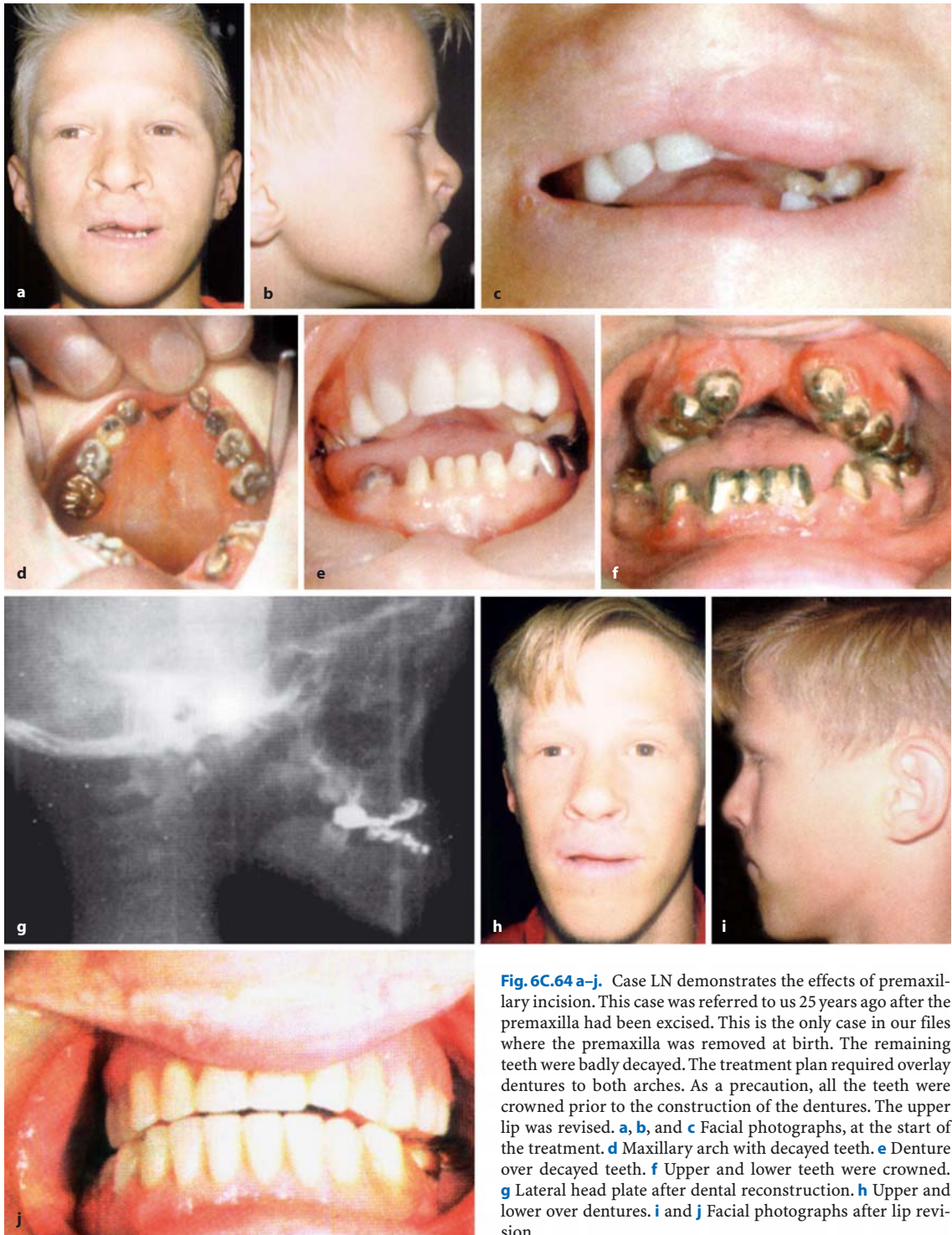


Fig. 6C.64 a-j. Case LN demonstrates the effects of premaxillary incision. This case was referred to us 25 years ago after the premaxilla had been excised. This is the only case in our files where the premaxilla was removed at birth. The remaining teeth were badly decayed. The treatment plan required overlay dentures to both arches. As a precaution, all the teeth were crowned prior to the construction of the dentures. The upper lip was revised. **a, b,** and **c** Facial photographs, at the start of the treatment. **d** Maxillary arch with decayed teeth. **e** Denture over decayed teeth. **f** Upper and lower teeth were crowned. **g** Lateral head plate after dental reconstruction. **h** Upper and lower over dentures. **i** and **j** Facial photographs after lip revision.



Fig. 6C.65. **a** Premaxillary excision. Removal of the premaxilla leads to a large oronasal opening with a retruded midface. Even at 1 year, 10 months of age, the upper lip is severely retruded. (courtesy of S Pruzansky.) **b** Premaxillary excision results in direct communication between the oral and nasal chambers

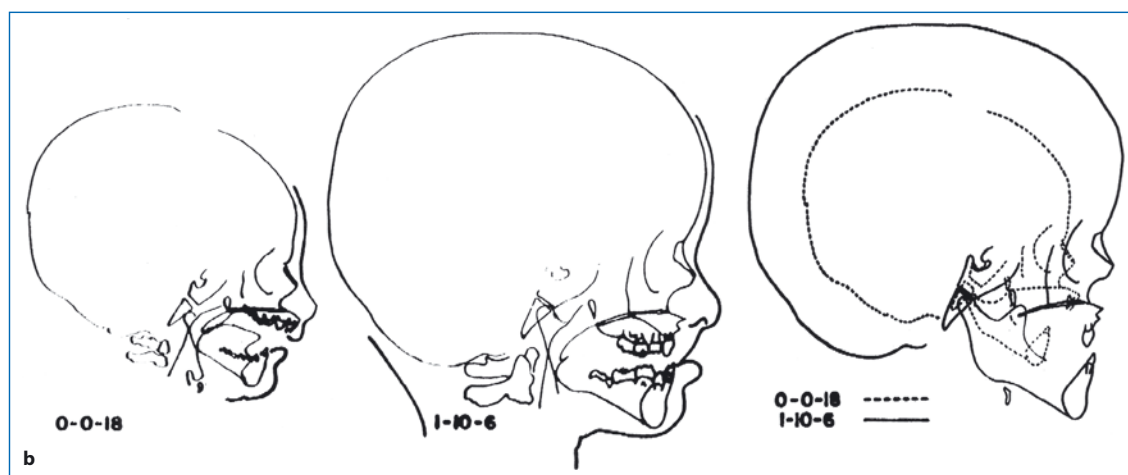




Fig. 6C.66 a-q. Case CH (II-64). This case demonstrates the need to advance the premaxilla after premaxillary surgical set-back. A chin augmentation was necessary to neutralize the effect of excessive upper facial growth. **a-i** An external elastic off a head bonnet was used to ventroflex the premaxilla prior to lip surgery. As a result of the continuous premaxillary overjet, at

4-9 it was surgically set back. The palatal cleft was closed at 5-6 using a modified von Langenbeck procedure. A secondary alveolar bone graft was placed at 10-5. The patient's family moved to a different state where the orthodontist unsuccessfully attempted to close the lateral incisor space by retracting the central incisors



Fig. 6C.66 a–q. (continued) **j** An anterior open bite was created. The patient returned to Miami. Treatment plan was changed to: (1) advance the premaxilla and open the lateral incisor space and (2) along with chin augmentation using autogenous bone from the inferior border of the symphysis. **k, l, and m** After

mandibular sagittal split surgery and orthodontic premaxillary advancement to recover lateral incisor spaces and advance the midface. A Hawley retainer with two false teeth were used to maintain the arch form

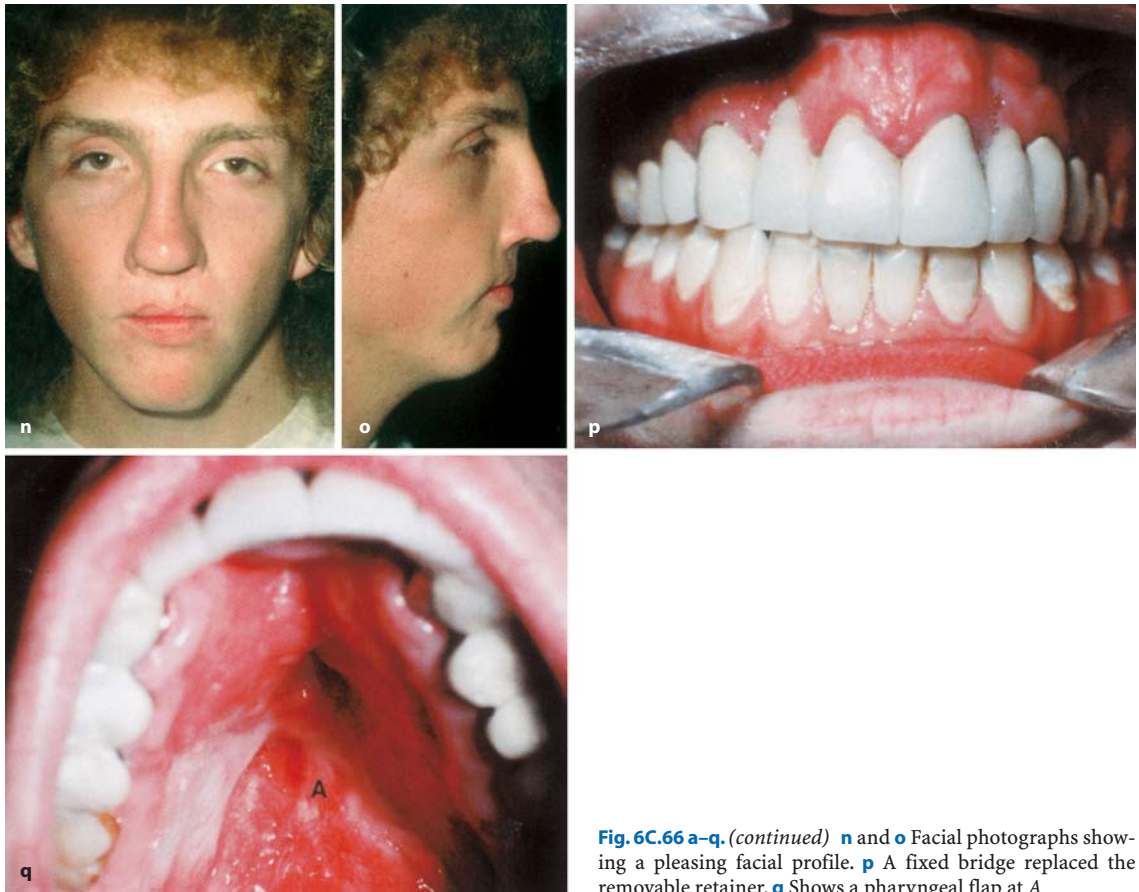


Fig. 6C.66 a-q. (continued) **n** and **o** Facial photographs showing a pleasing facial profile. **p** A fixed bridge replaced the removable retainer. **q** Shows a pharyngeal flap at A

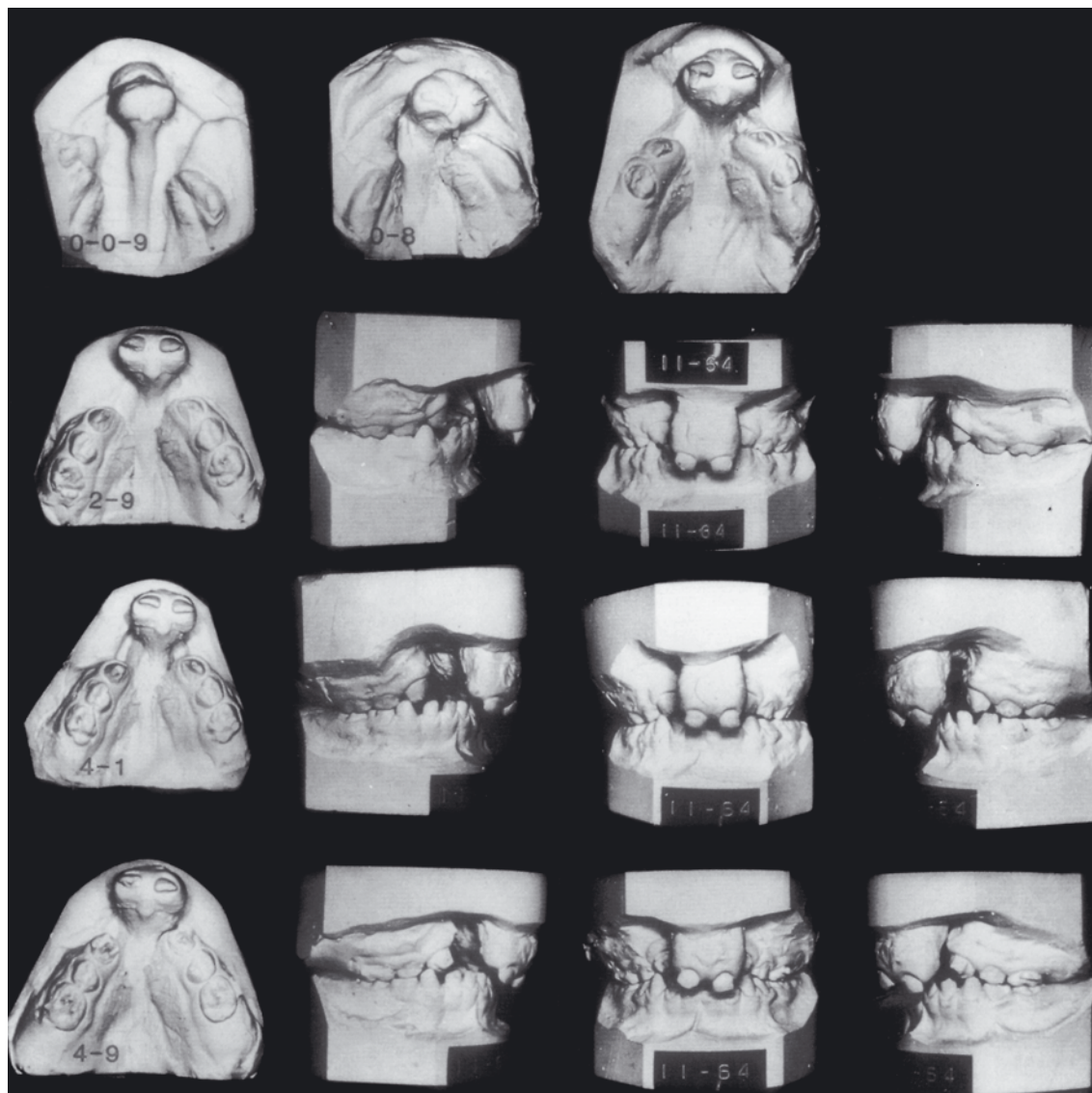


Fig. 6C.67. Case CH (II-64). Serial dental casts. 0-0-9 9 days of age. 0-8 Premaxillary ventroflexion after uniting the lip. The palatal segments have moved medially making contact with the vomer. The premaxilla has uprighted but the anterior cleft

space is slow in reducing. 2-9 Severe overbite and overjet. Mesioangular rotation of both palatal segments placed the deciduous cuspids in crossbite. 4-1 and 4-9 The anterior overbite and overjet has reduced spontaneously

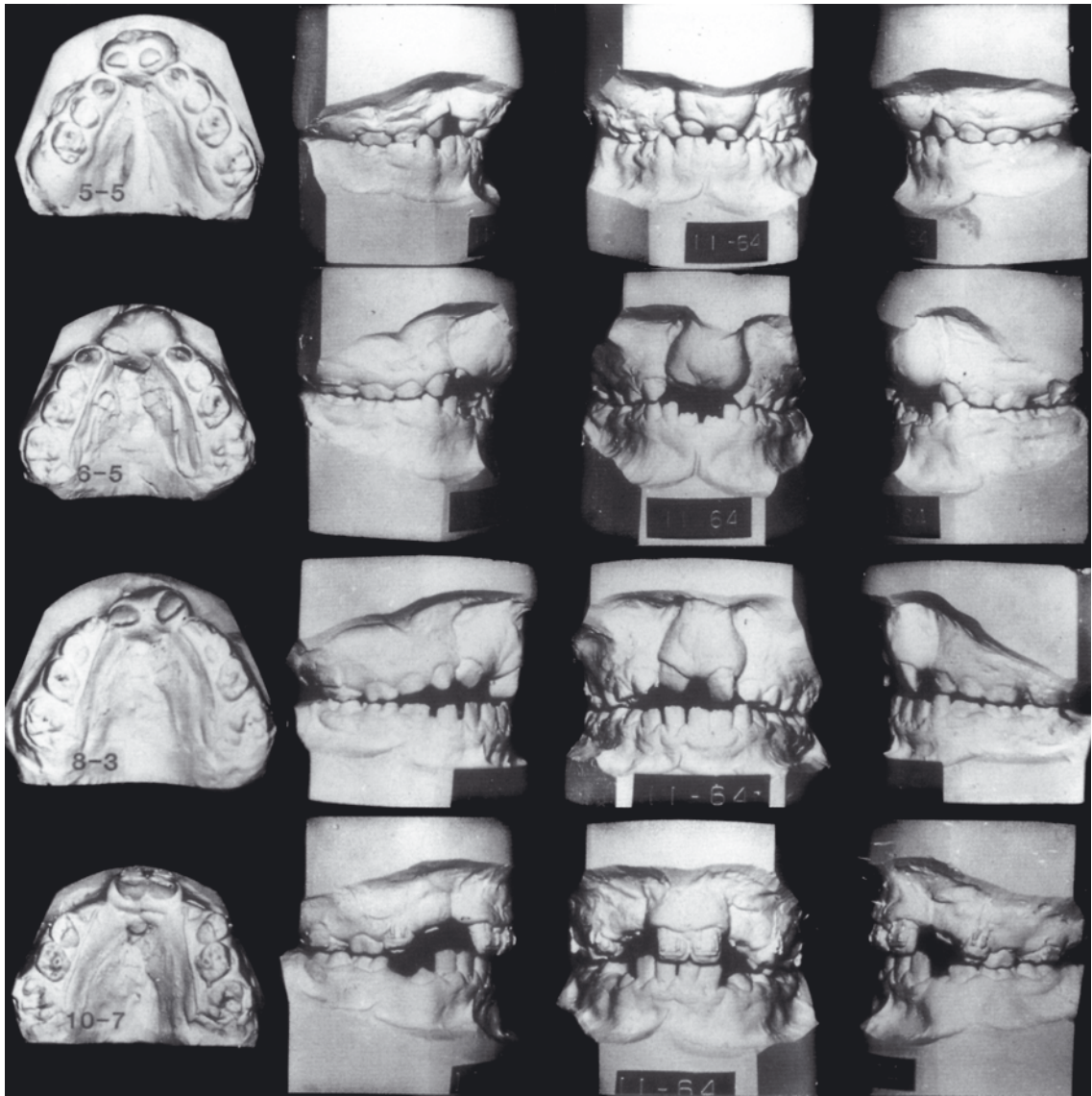


Fig. 6C.67. (continued) 4-11 The premaxilla was surgically set back. 5-5 The anterior open bite was eliminated. 6-5 The palate was expanded to correct the deciduous cuspid crossbite and to

allow the premaxilla to fit within the arch. 8-3, 10-7, and 11-5 Palatal retainer is used to hold the corrected arch form

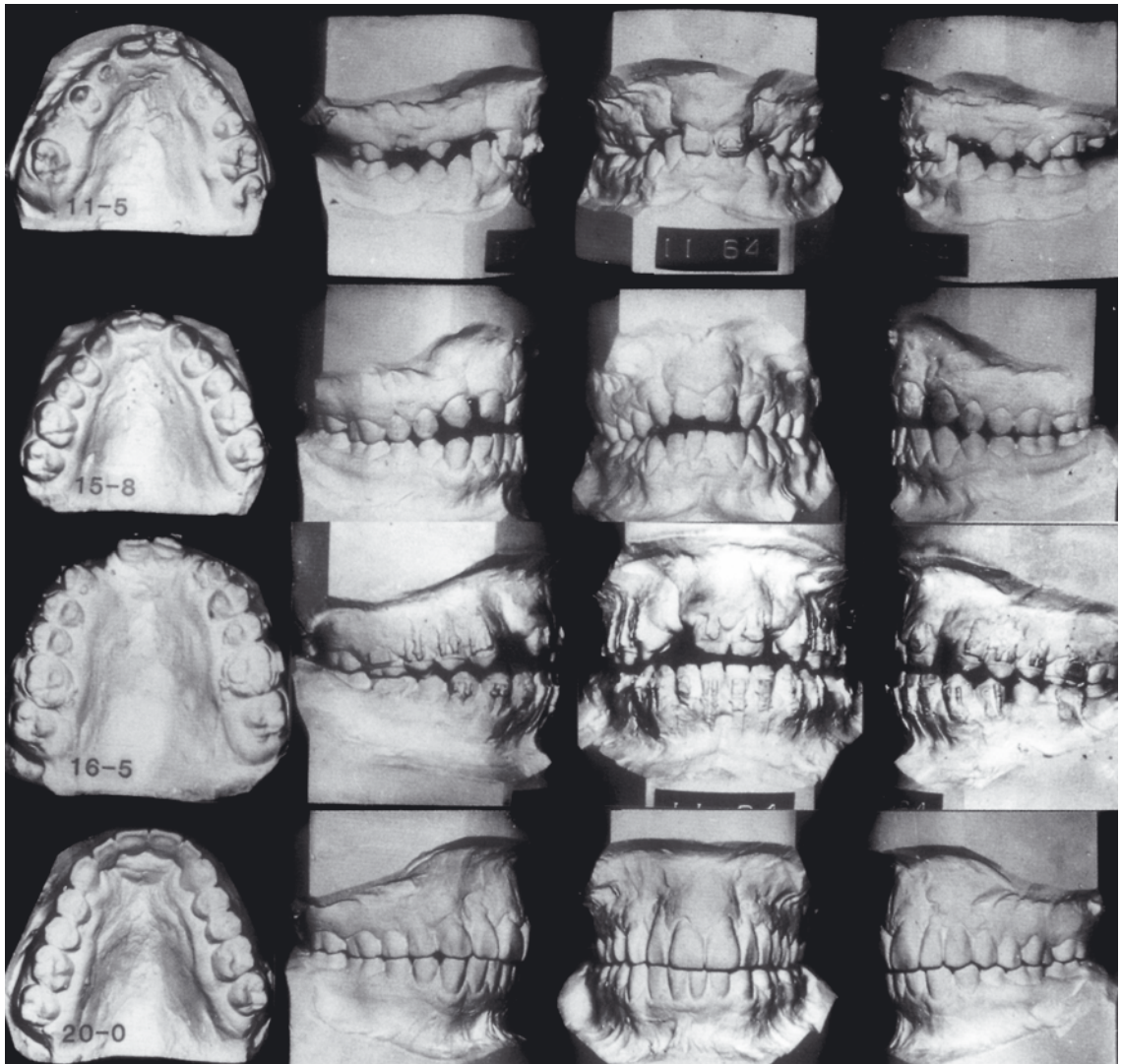


Fig. 6C.67. (continued) 15-8 An attempt to close the lateral incisor space orthodontically was unsuccessful. 16-5 Premaxilla advanced and lateral incisor spaces opened. 20 Fixed bridge replaces missing teeth and maintains the arch width and form

Fig. 6C.68. Case CH (II-64). Computer-generated palatal images drawn to scale. This series shows the gradual decrease in cleft space size from birth to 5-5 years. A posterior cleft space remained until 5-6. Alveolar cleft closed at 10-5. Comment: Hopefully, the research project currently under way will determine the best age, based on parameters such as the ratio of the palatal surface area to cleft space size, at which to close the posterior palatal cleft. The various facial growth studies presented in this chapter show great variation in palatal as well as facial growth patterns. Differential diagnosis to select the "best" time and surgical procedure to close the cleft defect – its size and the availability of tissue

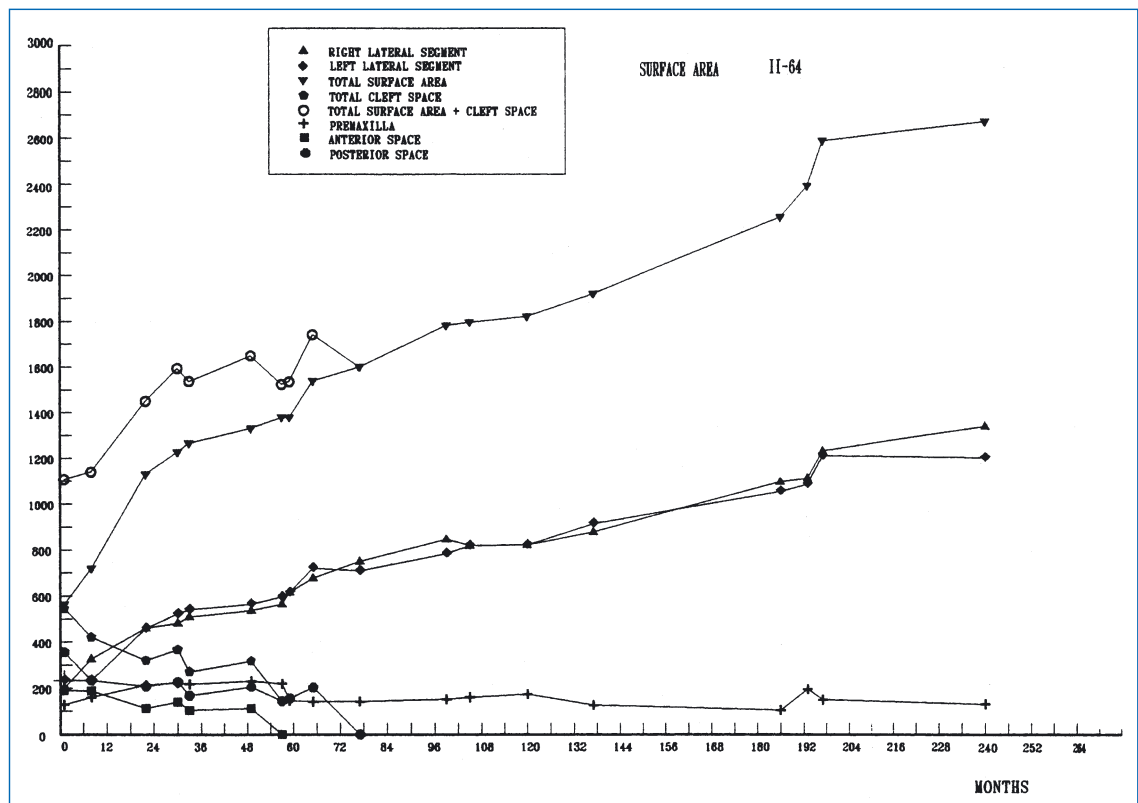
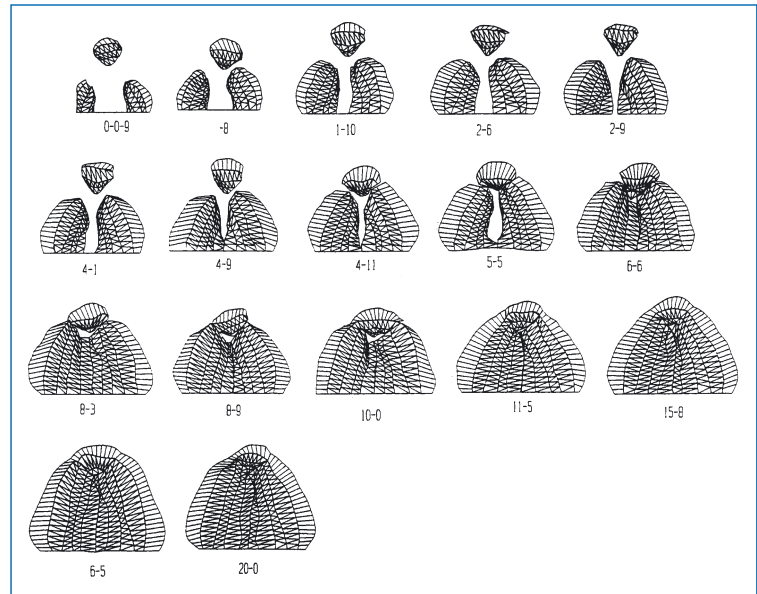


Fig. 6C.69. Case CH (II-64) A rapid increase in palatal growth as the posterior cleft space reduces at almost the same rate. The reduction in anterior cleft space proceeds at a lesser rate. Palatal growth rate after surgery is the same as it was before surgery

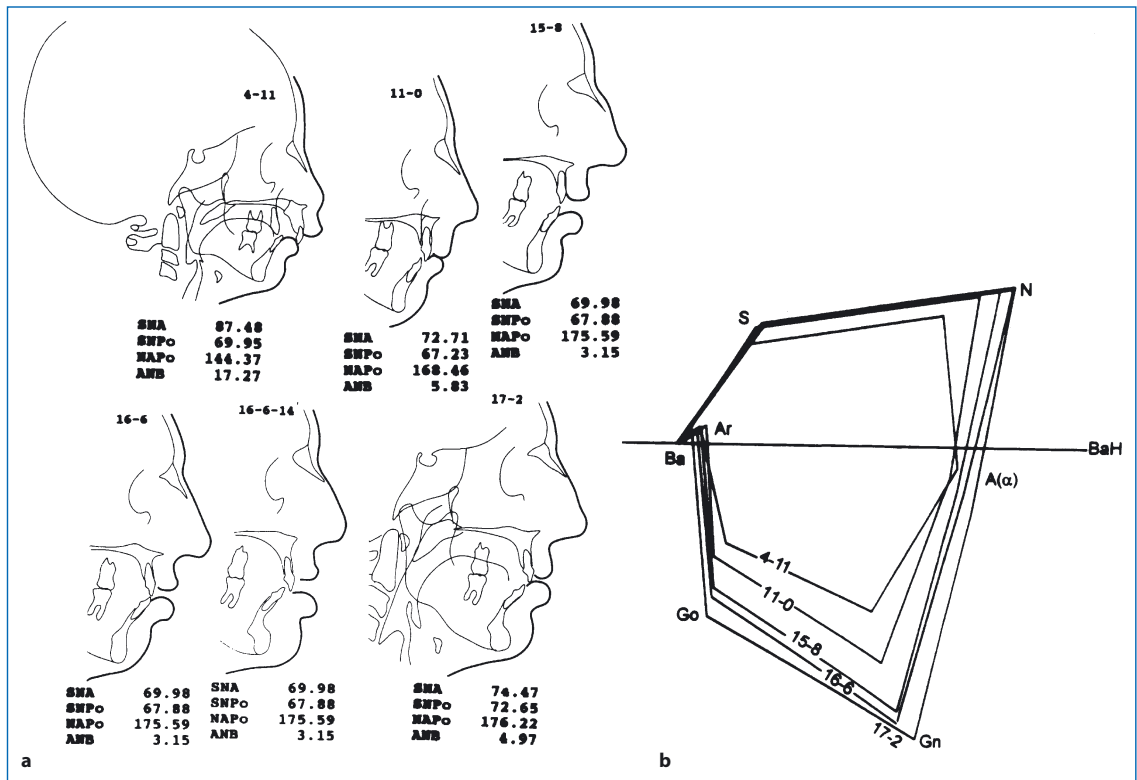


Fig. 6C.70 a,b. Case CH (IT-64). **a** Serial cephalometric tracings showing facial changes with a severely protruding maxilla at 4-11 and **b** superimposed polygons using the Basion Horizontal method (Coben). An excessive amount of growth occurred at the anterior cranial base (at N) and in the mandible mainly in the vertical dimension. With some minor midfacial growth the skeletal and soft tissue profile became more retrognathic and unaesthetic. Because of the prominent upper face, it was decided to improve facial aesthetics by advancing the premaxilla and to open the lateral incisor space. Notice the resulting larger incremental midfacial change resulting from the movement of A point between 16-6 and 17-2. Chin augmentation increased the mandibular prominence from an SNPo angle of 67.88° to 72.65°. With the lack of continued growth of the anterior cranial base between 16-6 and 17-2, the angle at facial convexity changed slightly from 175.59° to 176.22°. Comments:

The clinician should not evaluate the alignment of teeth only within the arch and occlusion without considering the effects of anterior tooth position on the developing facial profile. Retracting the maxillary central incisors along with A point to close the lateral incisor space made the midface appear very deficient and aesthetically unpleasant. Because of the large increment of anterior cranial base growth, it was necessary to advance both the midface and augment the chin. Therefore, when treatment planning, the protruding premaxilla at birth needs to be considered as part of the developing face and treatment options need to be left open until the facial growth pattern is identified and can be adjusted to its best advantage. It is important to emphasize that a mechanically or surgically repositioned premaxilla does not advance in the midface with growth as a non-retracted conservatively treated premaxilla will do



Fig. 6C.71 a–k. Case AO (BM-82) shows surgical repositioning of the premaxilla in an adult. This patient had been under orthodontic-surgical care elsewhere. The orthodontist was reluctant to orthodontically correct the severe premaxillary overbite and left it to the surgeon. Because of the patient's age, it was

decided that surgical premaxillary alignment was the treatment of choice. **a–k** Before and after surgery. The maxillary central incisors are stabilized with a Hawley retainer with two false teeth in the lateral incisor position

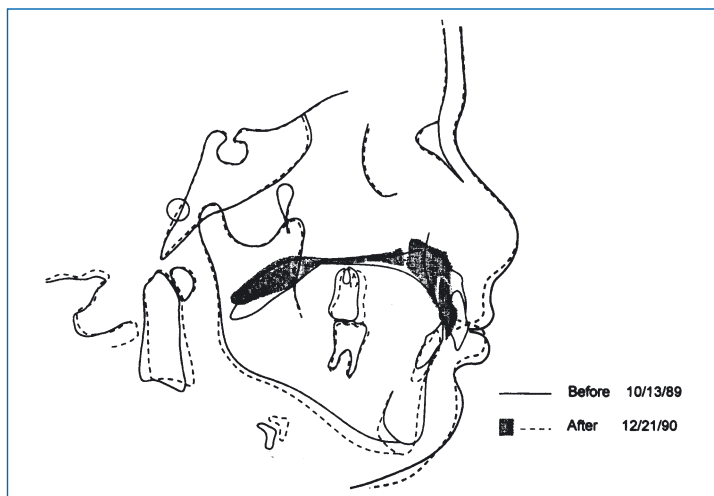


Fig. 6C.72. Case AO (BM-82). Lateral cephalometric tracings showing correction of the premaxillary overjet and overbite in an adult. Comment: When a severe premaxillary overbite exists in the mixed and early permanent dentition, in most cases it can be successfully treated with orthodontics. The surgical cut is made between the premaxilla and the vomer. The need to protect the integrity of the PVS is not crucial at this age. Rigid arch wire stabilizes the corrected premaxillary position during healing

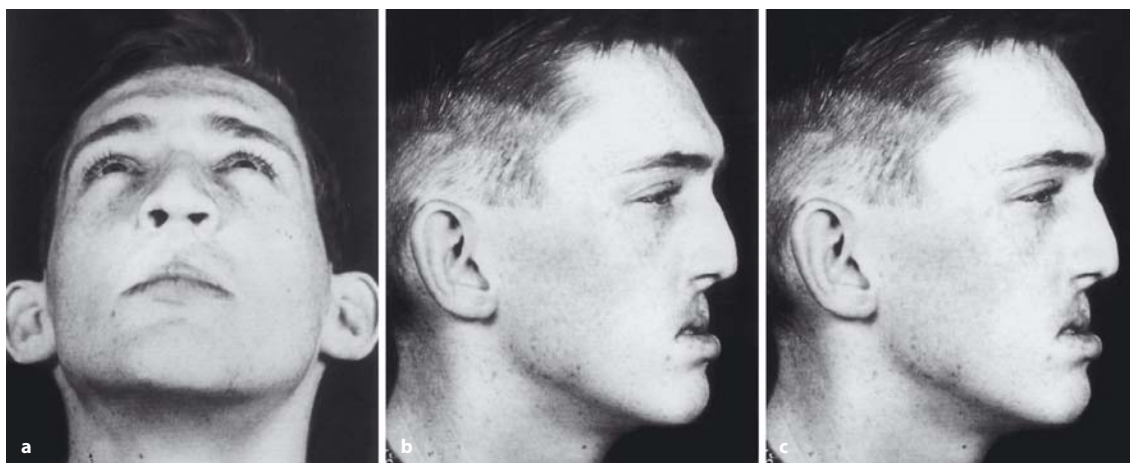


Fig. 6C.73 a-i. Case TK. Lip-Switch (Abbe Flap). The combination of a short columella and a short or long and tight upper lip requires secondary cleft lip surgery. Millard [68] strongly believes that, had the primary surgery been planned and executed properly, a lip-switch flap would most likely never be required. Unfortunately, there are occasions when the destruction of the lip landmarks and midfacial growth retardation require that tissue be brought in from outside and used to remove scars,

bring in muscle continuity, create a philtrum and even a bow, correct free border defects, and relieve tension. Millard suggests that the flap form the total central vertical length of the upper lip. It will then resemble the natural philtrum and can appear normal in spite of its scars. **a, b, and c** Repaired bilateral cleft lip and palate has resulted in a malformed, recessive midface with a scarred upper lip



Fig. 6C.73 a-i. (continued) **d, e,** and **f** The flap was taken from the middle of the lower lip and inserted in the total central portion of the upper lip. **g, h,** and **i** After lips were separated. The upper lip is symmetrical and shows a more normal looking vermillion. A dental prosthesis bumps the upper lip

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6D Isolated Cleft Palate

SAMUEL BERKOWITZ

In this cleft type (Fig. 6D.1), neither the lip nor the alveolar process is involved. The cleft may extend only through the soft palate or through the soft and hard palates, but it cannot exist in the hard palate alone because the fusion of the hard and soft palates proceeds from front to back (Figs. 6D.2, 6D.3).

The cleft may extend anteriorly as far forward as the nasopalatine foramen. The outline of the cleft space may be wide or narrow, long or short; any variation in geometric form can exist (Figs. 6D4–6D6; see Chap. 4, Facial and Palatal Growth).

Timing of surgical closure depends on the width and not the length of the cleft space. Hard palate clefts are frequently closed simultaneously with the soft palate cleft. In some instances, surgical closure of very wide hard palate clefts may need to be postponed until there is additional palatal growth, which may be as late as 5 or 6 years of age. An obturator with a pharyngeal extension (speech aid appliance) can be worn until the palate is closed (Figs. 6D7, 6D8).

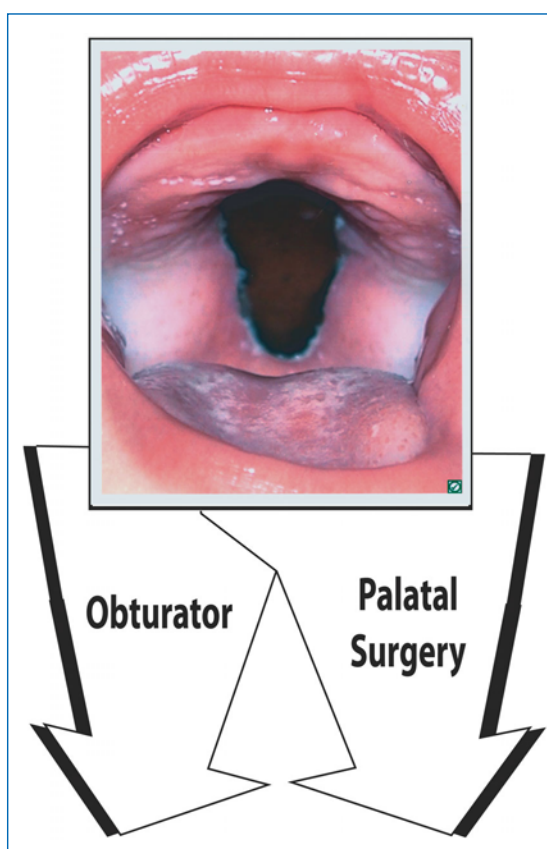


Fig. 6D.1. Isolated cleft palate



Fig. 6D.2. Isolated cleft palate. Serial casts demonstrate that, in some cases, good Class I occlusion can occur when closing the cleft space at 18 months of age, or even later, using a modified von Langenbeck surgical procedure. This occlusal result supports Berkowitz's contention that the "critical threshold level" for good palatal growth is determined by the size of the cleft space relative to the amount of available mucoperiosteal tissue.

This threshold, which has been determined to be a ratio of cleft size to palatal size medial to the alveolar ridges of 10% or less, is critical to the development of good arch form and occlusion. The threshold ratio, if exceeded by early palatal closure when the palatal size is relatively small compared to cleft size, will cause excessive palatal scar tissue resulting in midfacial growth inhibition.

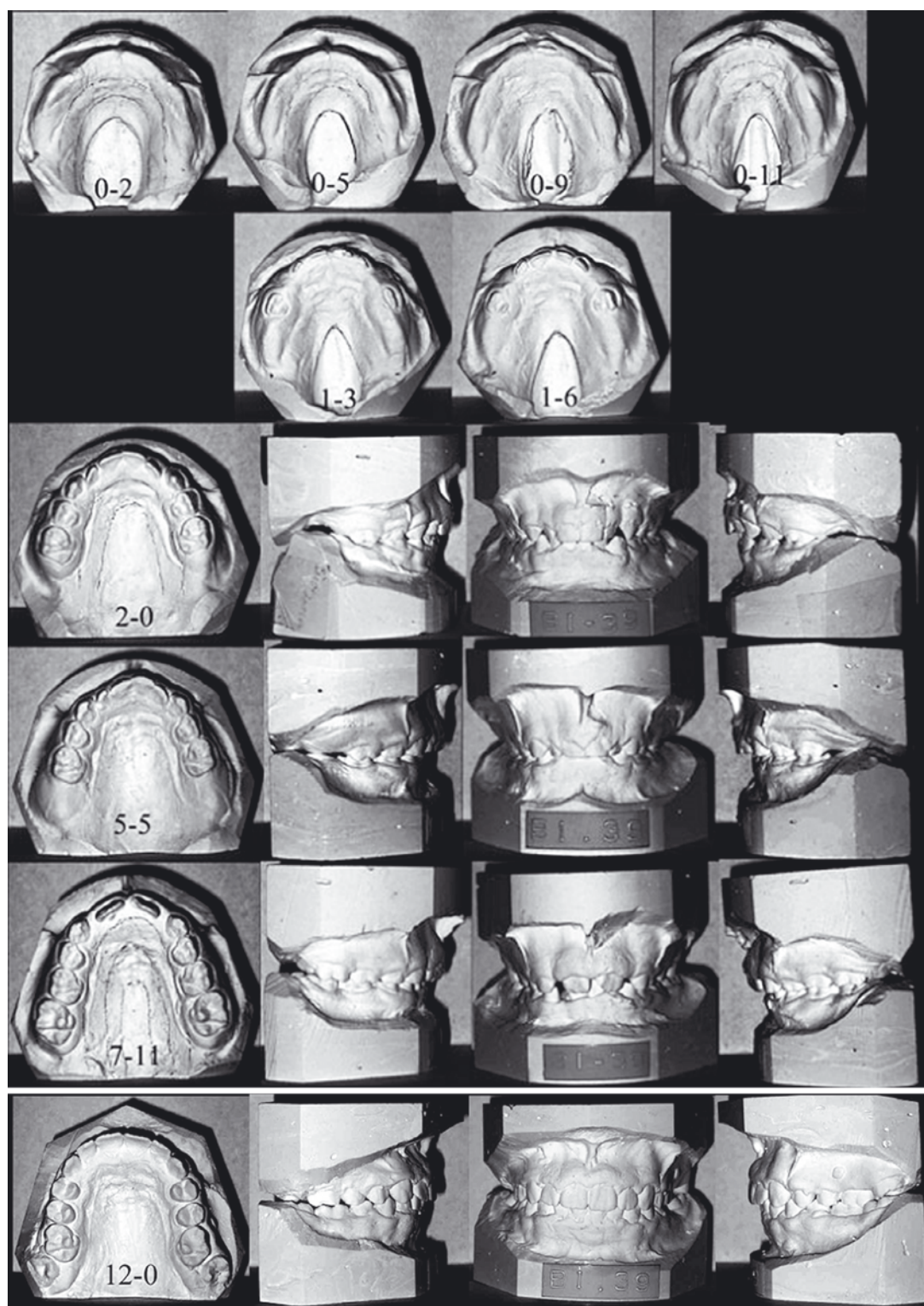


Fig. 6D.3. Case CR-BI 39. Patient with isolated cleft palate. Notice that the relative size of the cleft space diminishes as the palate grows and increases in size. The palate was closed with

the von Langenbeck at 18 months. Note the excellent occlusion which reflects on diminished palatal scarring. The patient has excellent speech

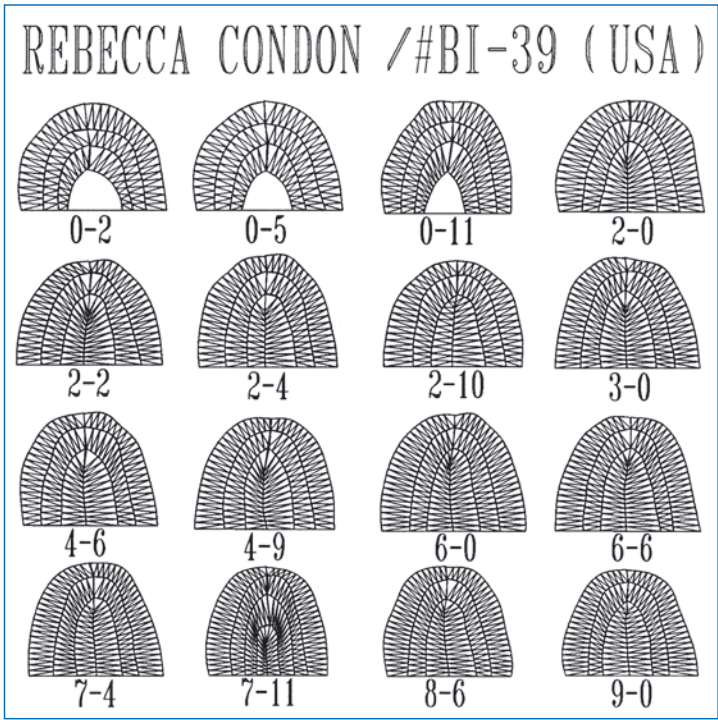


Fig. 6D.4. Superimposed computerized 3D tracings of isolated cleft palate. Isolated cleft palate. Superimposition horizontally on the rugae and registered vertically on the vomer line. This shows that most of the growth occurs posteriorly to accommodate the developing deciduous molars. Growth changes of the cleft space is highly variable. In some instances the cleft space can narrow or stay the same size. However in all instance the palate increases in size so the ratio of cleft space to surrounding palatal size reduces. In most cases time for surgery, determined by the ratio of 10%, is reached at 18–24 months. It can be earlier or even later in some cases

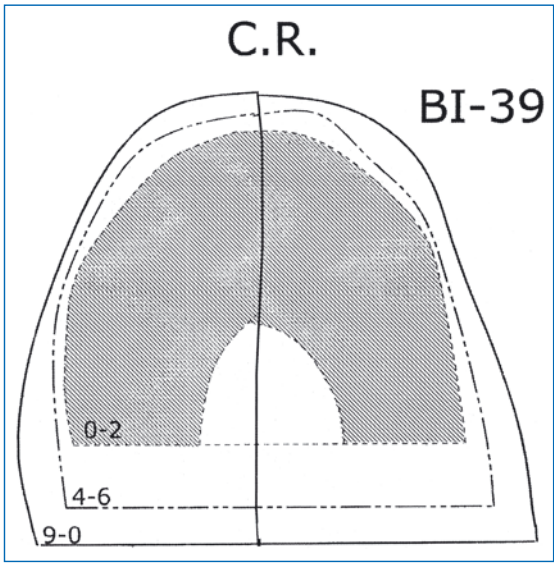


Fig. 6D.5. Case CR-BI 39

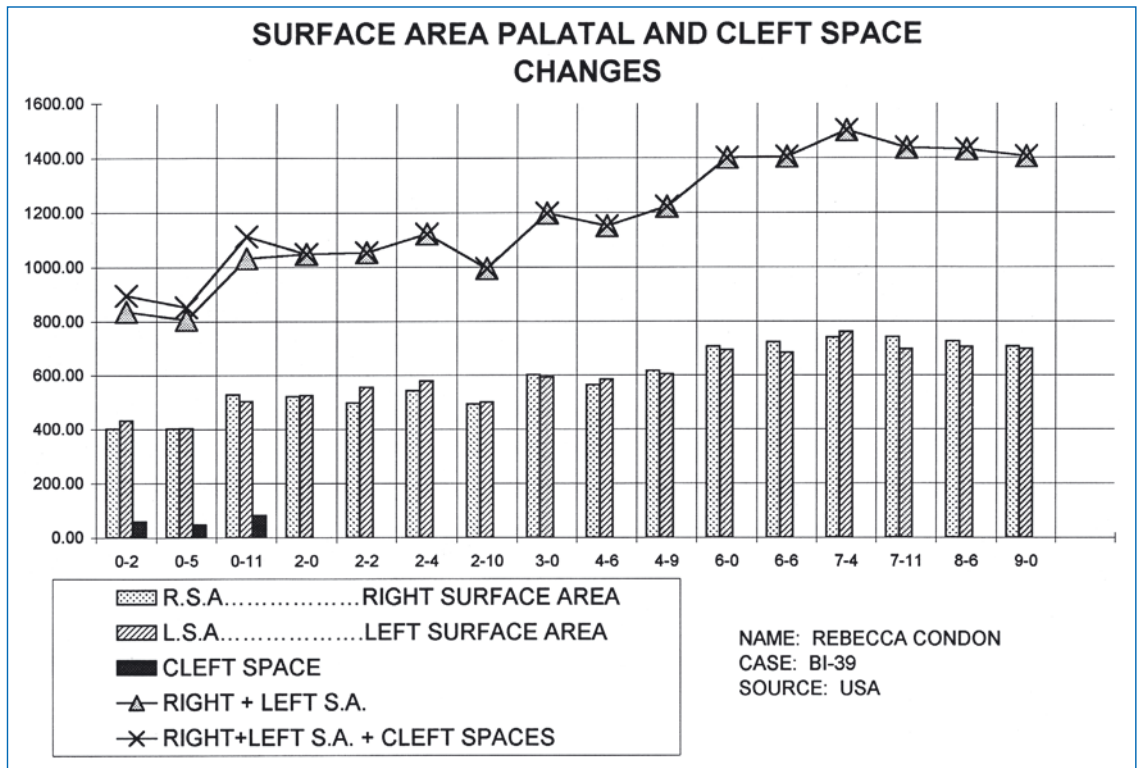


Fig. 6D.6. Serial size changes of the palate and cleft space

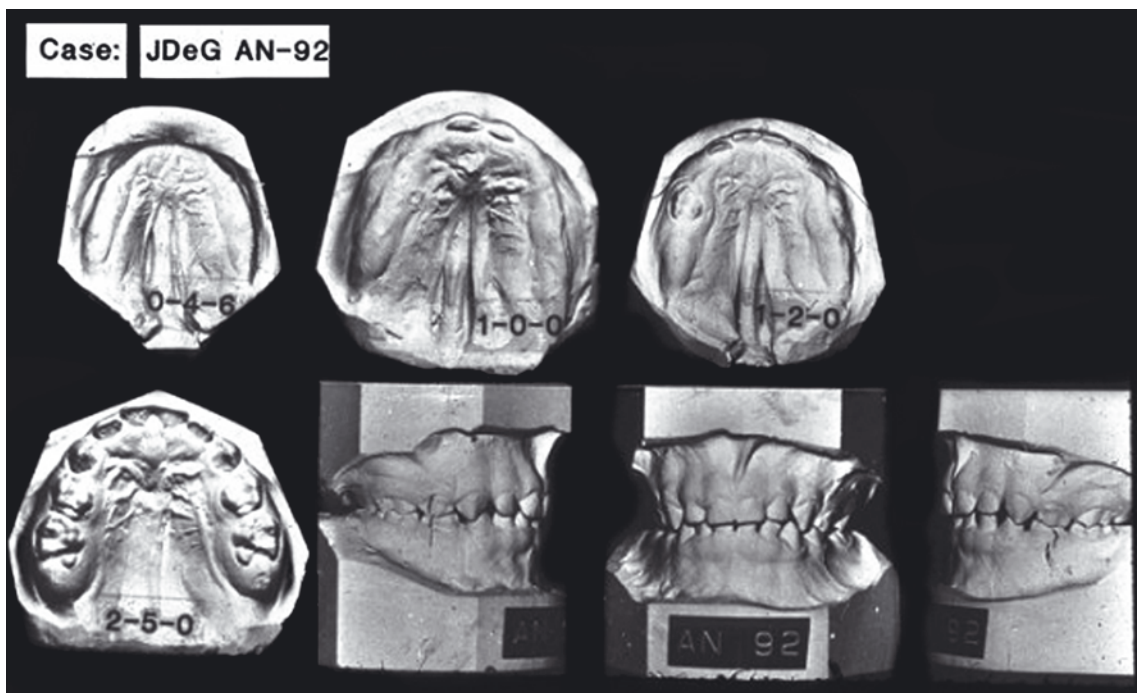


Fig. 6D.7. Case AN 92. Palatal closure at 1-5 using von Langenbeck procedure resulting in good occlusion

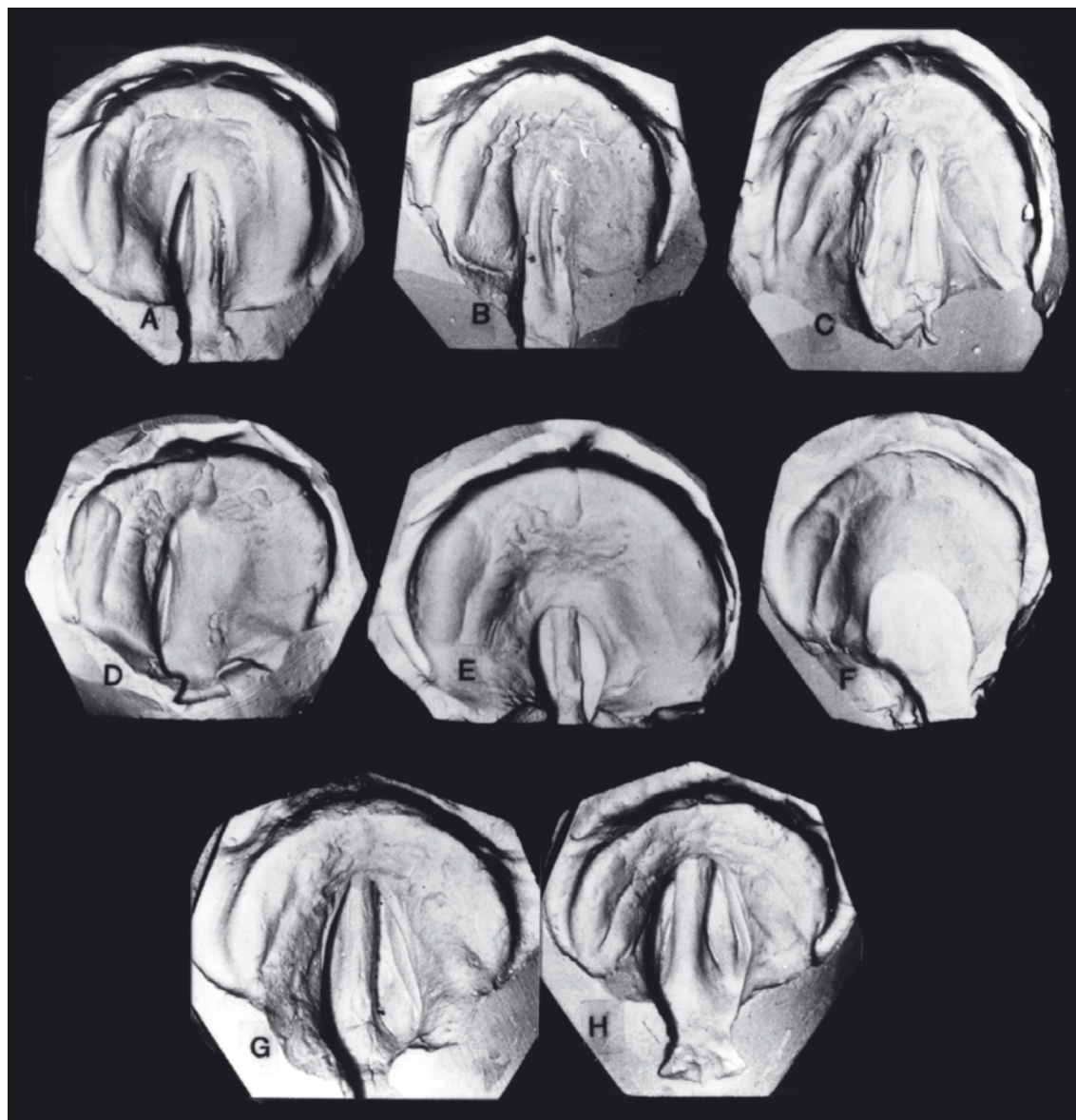


Fig. 6D.8. Variations in isolated cleft palate. The length and width of the cleft space is highly variable. The cleft extends anteriorly to various distances but not beyond the incisal canal. Note the relative size of the cleft palate space which reflects

differences in the degree of osteogenic deficiency. In some instances, the cleft space diminishes with time; however, there are occasions when the cleft's relative size remains the same

6E Submucous Cleft Palate

SAMUEL BERKOWITZ



Fig. 6E.1. This cleft is characterized by a bifid uvula, lack of muscle continuity across the soft palate, and a pink zone of mucosa (zona pellucida) across the cleft in the hard palate.

A palpable notch in the posterior border of the hard palate is always indicative of the presence of a cleft. Not all patients with submucous cleft have VPI

7

Lip Pits; Orthodontic Treatment, Dentition and Occlusion; Associated Skeletal Structures

SAMUEL BERKOWITZ

7.1 Lip Pits

7.1.1 Pits of the Lower Lip in Cleft Lip and/or Palate – Genetic Considerations

Pits of the lower lip such as fistulas of lower lip, paramedian sinuses of lower lip, humps of lower lip, or labial cysts are a very rare congenital malformation, first described by Demarquay in 1845 [1].

This minimally deforming anomaly is remarkable chiefly for its association with facial clefts. The fact that clefts that occur with lip pits seem to run stronger in families than clefts without lip pits has attracted the attention of professionals dealing with cleft patients (Figs. 7.1–7.5).

7.1.2 Frequency

No survey of lip pits has been carried out among the general population; hence the frequency of this rare anomaly has been only roughly estimated from its incidence in hospital records.

Assuming that 70% [2] to 80% [3] of patients with pits of the lower lip have associated cleft lip and/or palate, and that the frequency of clefts is 1:650 (one in every 650 births), it can be estimated that the frequency of lip pits among the general population is about 1:75,000–1:100,000 (one in every 75,000 to 100,000 births).

Fig. 7.1. Lower lip pits in a child with a bilateral cleft lip and palate. **a** Before lip surgery. **b** After lip surgery at 6 months





Fig. 7.2. **a** Pits in the upper and lower lips in a 20-year-old with a bilateral cleft lip and palate. **b** Close-up view



Fig. 7.3. Lip pits in the upper and lower lips in a bilateral cleft lip and palate

7.1.3 Morphology

Fistulas of the lower lip usually appear as two pits or humps on the vermillion portion of the lower lip, generally equidistant from the midline. Various kinds of asymmetry may be observed with regard to the midline, or one pit may be positioned more orally. Some pits are mere depressions; others are channels 10–15 mm deep with openings at the top of nipple-like elevations. Some secrete small amounts of viscous saliva but most are asymptomatic.

In exceptional cases, only one pit is present, which may be located either centrally or on one side or the other of the midline of the lower lip. Some cases of single lip pit have occurred in families with members with double lower-lip pits. It can be assumed that a single pit is not a distinct entity but rather an incomplete expression of the trait. On the other hand, the rarely described fistulas of the upper lip [4, 5] have not shown any inheritance pattern.

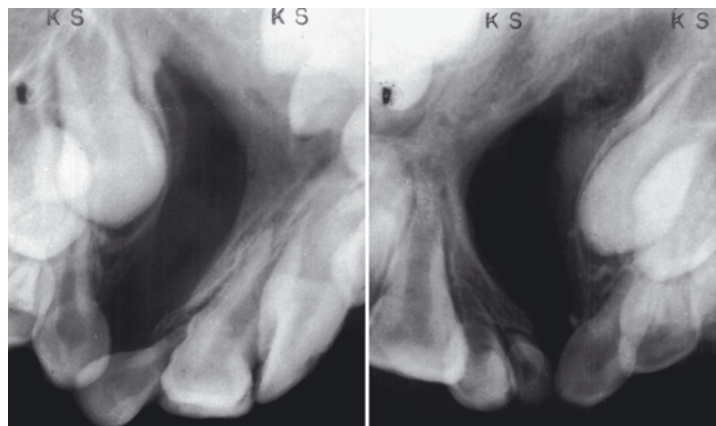


Fig. 7.4. Radiograph of a premaxilla showing two central and one lateral incisors (*left*) and twined lateral incisors and one malformed central incisor (*right*)



Fig. 7.5. Missing and malformed anterior teeth. **a** A very small premaxilla with one deciduous incisor but no permanent tooth buds. **b** Malformed deciduous maxillary anterior teeth and missing left deciduous anterior teeth

Commissural or angular lip pits – small, usually asymmetrical channels located at the lip angles – are also distinct entities with a much higher incidence and different embryology [2, 6–10].

7.1.4 Association with Other Malformations

In addition to their strikingly common association with cleft lip and/or palate, pits of the lower lip have been noted in association with other malformations. Gorlin and Pindborg [2] have found cases of lip pits along with anomalies of extremities, popliteal pterygia, and anomalies of the genitourinary system. The association with cleft lip and/or palate may well form a distinct new syndrome.

A review of the literature suggests that a variety of other anomalies may be associated with lip pits. Among them: syndactyly of the hands together with cleft lip and palate [4, 11]; mental retardation and cleft, type not specified [12]; ankyloglossia and cleft lip and palate [3]; polythelia [14]; symblepharon and cleft lip and palate [15]; and ankyloblepharon, adhesion between the maxilla and mandible, and cleft uvula [16]. In two cases of the orofacial digital syndrome, pits were observed by [17].

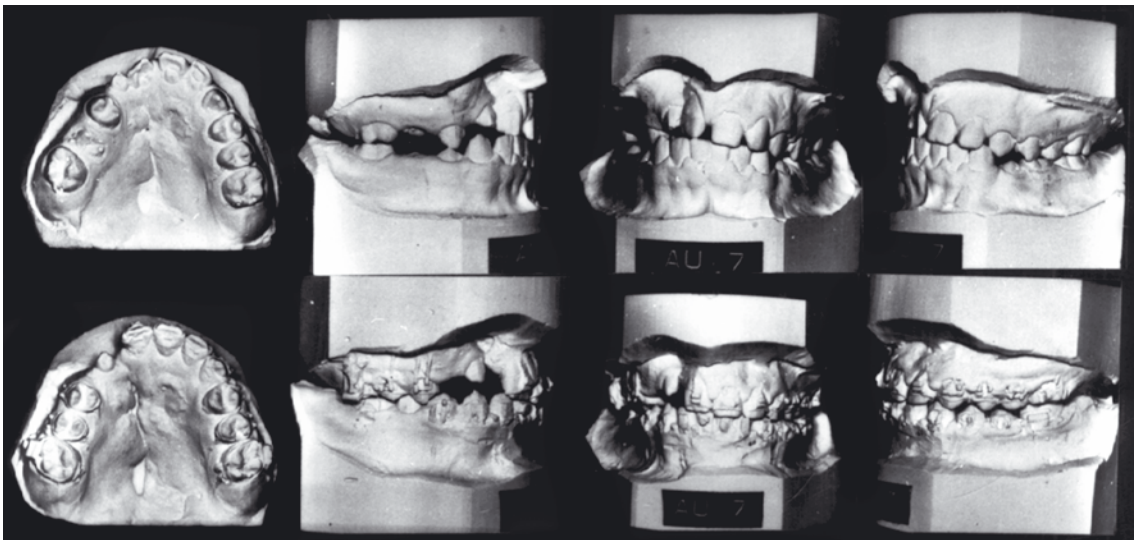


Fig. 7.6. Palatal expansion increases nasal width. At 12 years of age buccal crossbite correction of the cleft segment was performed by palatal expansion. Crossbite correction is orthopedic in that the cleft bony segment is moved laterally, widening the

nasal chamber on that side. *Top:* Right buccal segment is in crossbite. *Bottom:* After expansion: Palatal fistulae are exposed with the separation of the overlapped palatal segments

7.1.5 Inheritance

In most cases of lip pits described during the last 120 years, a marked hereditary pattern was observed. Although all authors have excluded autosomal recessive inheritance or X-linked inheritance, there is no uniform opinion concerning whether or not the condition is due to a single autosomal dominant pleiotropic gene.

Fogh-Andersen [18, 19] was the first to clearly point out that the inheritance of clefts in the families with a history of lip pits is of a different character than in families where no lip pits occur. According to Fogh-Andersen [18, 19] the role of genetic factors in families with lip pits is much more pronounced, and both genetically different types of clefts (cleft lip or cleft lip and palate and isolated cleft palates) commonly are found within a single family. Fogh-Andersen also stated that, in families in which fistulae of lower lip occur as a dominant hereditary character, there are some cases of cleft lip and cleft palate alone. Possibly it may be explained as the result of coupling of neighboring genes.

Van der Woude [3], in a careful study of five pedigrees with clefts and lip pits, found that the combination of pits and clefts is based on a single dominant gene of variable expressivity. She agreed with other authors [12] that a mildly affected individual can pass the trait on in a very severe form, and a severely affected individual can pass the trait on in mild form. The sex of the individual is not a factor in passing on this anomaly. No sex limitation or preference exists.

Patients with clefts but without lip pits (or their parents) often ask for genetic advice. In most of these cases, only a small risk of cleft lip (less than 10%) is indicated for the next child. But the counseling situation is significantly changed when associated lip pits are found. In this instance, all clefts in the family are considered part of the syndrome, and risk figures for clefts are remarkably higher.

7.1.6 Evidence of Heterogeneity

It is also clear that the risk of a cleft occurring in a child is significantly higher when the parent has lip pits and a cleft than when the parent has lip pits only. Two alternative explanations for this heterogeneity between families can be considered: (1) The development of clefts in persons carrying a "lip pit" major gene may be influenced by modifying genes at other loci; and (2) in some families, a mutant allele may produce lip pits with only occasional clefts; whereas in other families a different mutant allele (at the same or a different locus) may frequently lead to clefts in addition to lip pits. Thus far, efforts to use the data to support one or the other of these hypotheses have been unsuccessful. Cervenka et al. [20] reported data from 66 individuals with lip pits in his study and 446 cases with lip pits from the literature with known sex to establishing a 1:1 sex ratio. The frequency of the syndrome was estimated as 1:75,000 to 1:100,000 in the white population.

Cervenka [20] further states that family histories can be explained adequately on the basis of autosomal dominant inheritance with variable expressivity of the trait. Penetrance is high, estimated at 80%. Pits show up more frequently than clefts, and there is a significant association between the types of clefts in parents and their children. Possibly the development of clefts in this syndrome is influenced by modifying genes or by different mutant alleles with a predilection for the different types of cleft.

7.2 Orthodontic Treatment, Dentition and Occlusion

7.2.1 Crossbite Correction (Figs. 7.7–7.12)

Bergland and Sidhu [21] advocated postponing orthodontic treatment until complete eruption of the permanent anterior teeth. Segmental alignment can then be corrected with simultaneous manipulation of the anterior teeth. We believe it is best to start maxil-

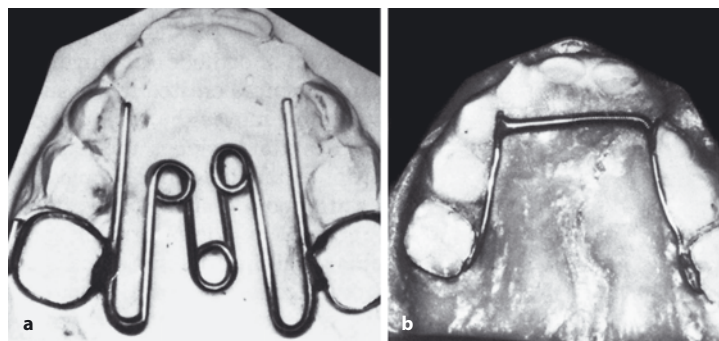


Fig. 7.7. **a** Fixed palatal helix expander used in deciduous dentition with or without an anterior finger spring. **b** Fixed "Arnold" expander using a compressed open coil spring to create an expansion force

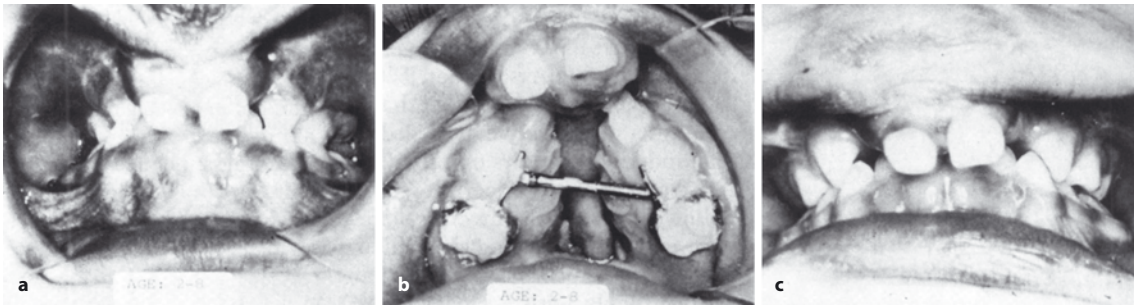
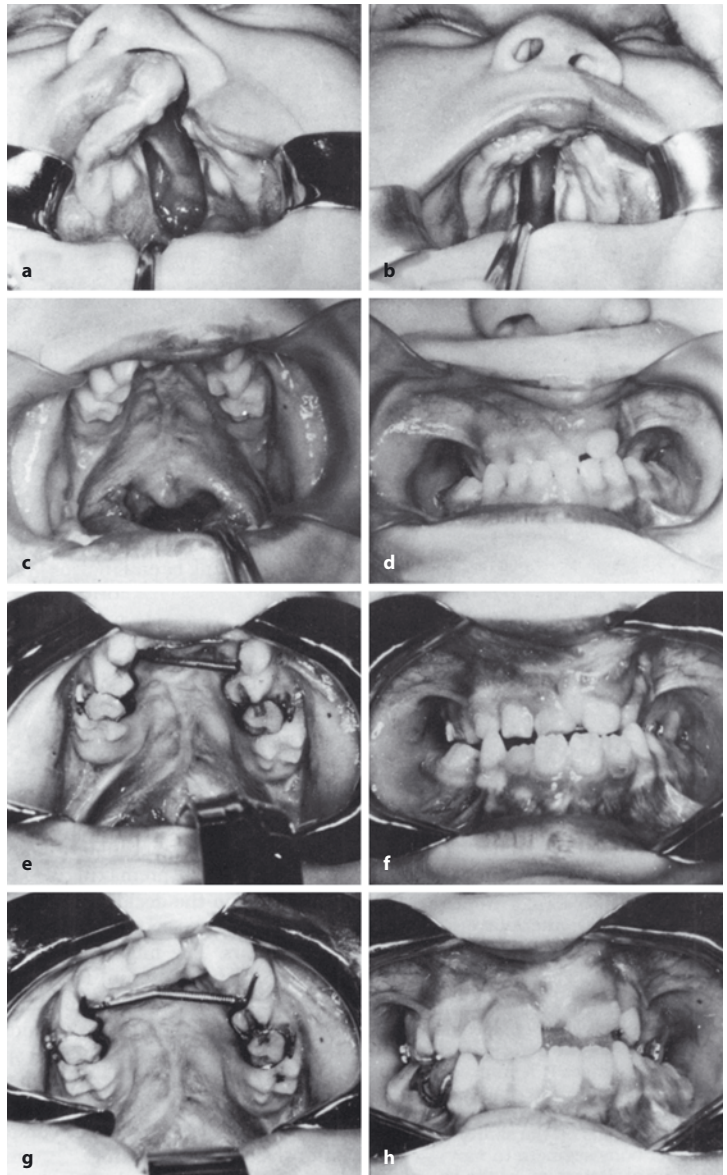


Fig. 7.8 a–c. Crossbite correction for bilateral cleft lip. **a** Age 2 years 8 months. Note the bilateral crossbite. **b** Arnold expander in place. **c** Crossbite correction after 3 months brought on by the outward movement of the lateral palatal segments

Fig. 7.9 a–h. Correction of anterior and posterior crossbite in the deciduous and mixed dentition in a severely scarred palate. **a** Complete unilateral cleft lip and palate, unoperated. **b** Palatal segments in contact. **c** Palatal cleft closed at 6 months of age; von Langenbeck procedure was used to close a very wide cleft space. **d** Bilateral buccal and anterior crossbite. **e** Arnold expander is in place. **f** Crossbite is still present due to excessive scar tissue. **g** Expander is fully extended in the early mixed dentition. **h** Anterior crossbite was corrected, but the buccal crossbite is still present. Comments: The severely scarred palate prevents the lateral movement of the medially positioned palatal segments. Scars can only be stretched a slight amount, and if that amount is exceeded the bony segments will not move and instead the teeth will respond to the orthodontic forces. Should the teeth be tipped outwardly they will have to be permanently retained by a bridge or by teeth splinting



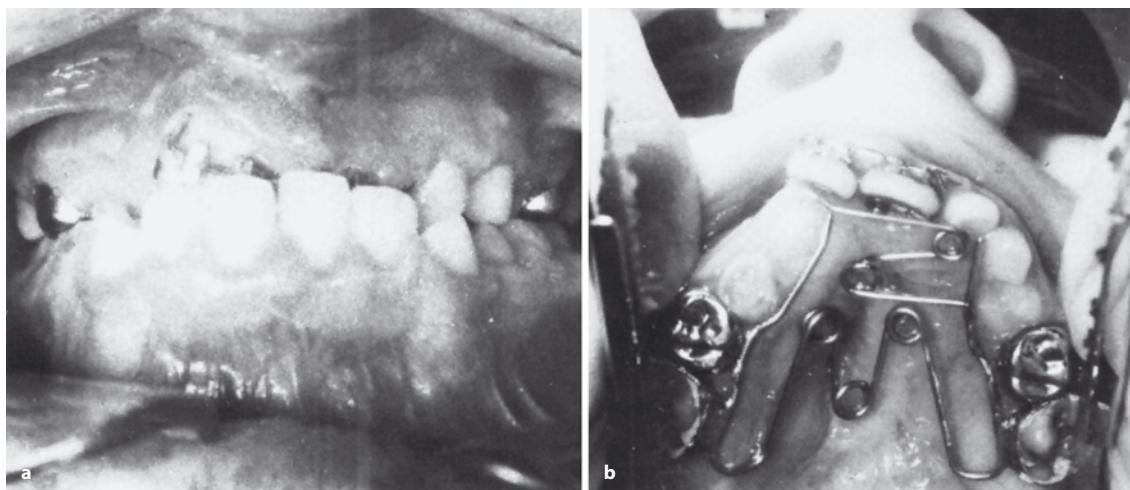


Fig. 7.10 a,b. Anterior and buccal crossbite correction the mixed dentition. **a** Anterior crossbite was due to palatally displaced deciduous teeth with premaxillary segment and not to growth deficiency. The buccal teeth are in crossbite as a result of the lesser palatal segment being palatally displaced. This picture has the maxillary anterior teeth in crossbite; the lower anterior teeth are shown while the upper teeth are hidden.

b A fixed palatal expander with finger springs is used to expand the arch and advance the anterior teeth. In most instances it is unnecessary to disocclude the anterior teeth to move them forward. Buccal expansion depends on the ability of the lesser bony segment to move outward. Retention of the bony correction is essential

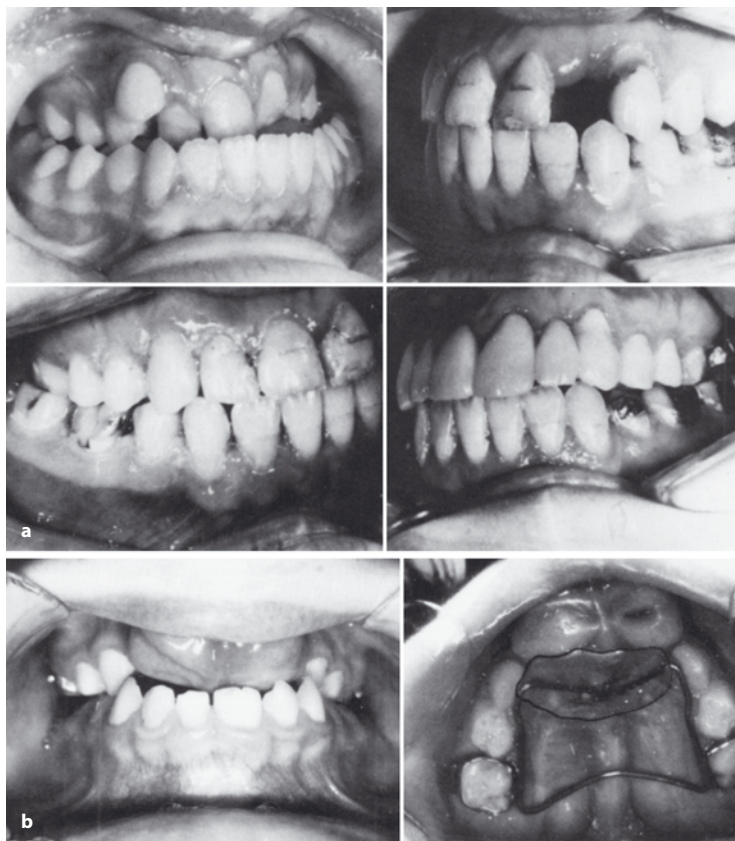


Fig. 7.11. a Orthodontic correction of an anterior and posterior crossbite in the permanent dentition. Anterior dental crossbite does not necessarily mean that the maxilla is anteroposteriorly deficient in size and requires a LeFort I advancement. In this case the maxillary dentition was advanced orthodontically with the missing lateral incisor space opened to achieve interarch congruency. The expansion was maintained by complete arch splinting. **b** A severely ventroflexed premaxilla was uprighted during the deciduous dentition. In order to maintain the correction after the deciduous anterior teeth are lost a fixed palatal retainer with an acrylic button is placed on the premaxilla's palatal incline

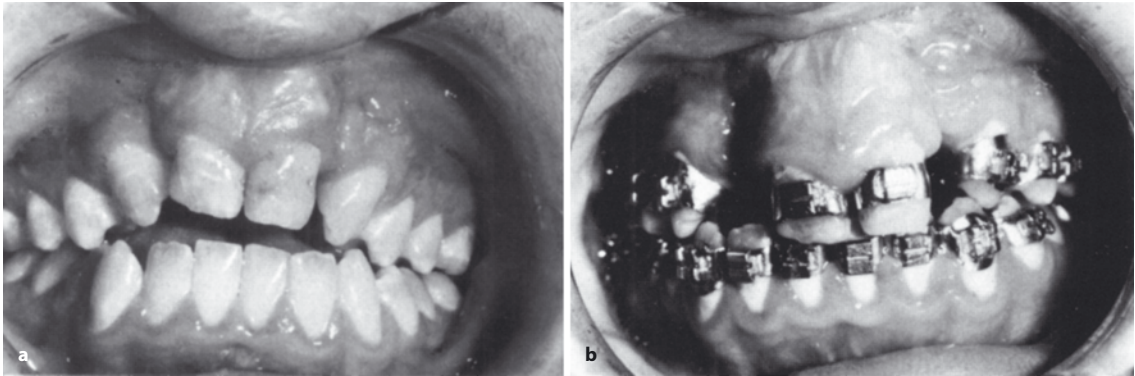


Fig. 7.12 a, b. Repaired bilateral cleft lip and palate. An anterior openbite and retruded premaxilla resulting from inadequate orthodontic and surgical planning. **a** A slight anterior openbite was present at 8 years of age at which time a secondary alveolar bone graft was performed. The orthodontist attempted to close the lateral incisor spaces by bringing the cuspids mesially, while retracting the central incisors. This created a more severe openbite and an anterior crossbite. **b** The orthodontic mechanics were reversed. The lateral incisor spaces were opened and the

central incisors were advanced into an ideal overbite and overjet relationship. One of the bone grafts had to be redone. Comments: When the buccal occlusion is class I it is always better to keep the lateral incisor space open in both bilateral and unilateral cleft cases. There are occasions when a mandibular central incisor will need to be extracted in order to avoid flaring the maxillary central incisors and to obtain a proper occlusion. An anterior fixed bridge will stabilize the arch and replace the missing maxillary lateral incisors

lary arch expansion when the deciduous dentition is completely erupted and when the children can be easily managed. A three-phase treatment is followed: (1) Buccal crossbite is corrected at 4–6 years; (2) anterior teeth are aligned at 8–9 years; and (3) final orthodontics is utilized at 11-plus years.

Crossbite correction by moving palatal segments laterally in the presence of extensive palatal scarring is difficult and often unsuccessful. Extensive mucoperiosteal undermining, leaving wide denuded palatal bone, was necessary to close the wide palatal cleft at 6 months of age. Uniting the lip moved the palatal segments together (molding action) into good arch approximation. Palatal scar contracture moved the buccal segments farther medially, placing the buccal teeth in crossbite. The anterior dental crossbite does not necessarily reflect diminished anteroposterior maxillary growth but, rather, malposition of the premaxillary teeth of the greater segment.

7.2.1.1 Unilateral Cleft Lip and Palate

Deciduous and Mixed Dentition. When the smaller segment's alveolar process is contained within the premaxillary alveolar segment of the larger segment, a dental crossbite occurs between the maxillary and mandibular teeth. Dental dysplasia (eruption of a tooth out of position) may or may not coexist with segmental dislocation. A simple crossbite of one tooth

may be due to its malposition rather than to the palatal segment's collapse. The most frequent crossbite is brought on by the mesioangular rotation of the lesser segment rather than by ectopic tooth eruption. Total buccal crossbite is seen less frequently but is generally present when the palatal tissue is scarred. In either case, the same helix type of palatal expander can be utilized and correction achieved within 2 to 4 months. Anterior movement of the deciduous central incisors often requires lingual undercuts either by orthodontic bands with lugs or by direct lingual shelf bonding to stabilize the anterior activated palatal finger springs.

A fixed palatal retainer can hold the correction until the second stage of orthodontic therapy is initiated. There is no way to predict the return of a crossbite in the absence of permanent retention. Bone grafting across the alveolar cleft does not guarantee retention of the corrected arch form.

Permanent Dentition. In Class I and Class III cases it is usually preferable to open the missing lateral incisor space with the expectation of utilizing a fixed bridge to stabilize the corrected arch form and replace the missing tooth. In Class II cases it may be possible to encourage the cuspid to erupt through the alveolar bone graft in the lateral incisor space, which eliminates the need for extensive orthodontics and bridge-work. The cuspid may need to be splinted to the central incisor in order to maintain the corrected arch form.

7.2.1.2 Bilateral Cleft Lip and Palate

First Stage. Between the ages of 4 to 6 years, the premaxilla is usually ventroflexed and overlaps one or both lateral palatal segments, which may be in partial or complete buccal crossbite. As in unilateral cases, treatment of bilateral cleft lip/palate necessitates moving bony segments into the surrounding muscle ring. If the premaxilla is to be moved forward, bands with lingual lugs are placed on the deciduous central incisors. A fixed helix-type palatal expander with anterior-activated finger springs is cemented to the second deciduous molars. The finger springs are positioned under the central incisor-banded lugs for retention. The premaxilla is uprighted prior to correcting the buccal crossbite. The anterior and buccal crossbite correction can be completed within 6 months. As the premaxilla and lateral palatal segments are moved outward, the anterior cleft space is uncovered. A fixed palatal retainer with an acrylic anterior extension to cover the anterior cleft space is placed and kept in position until the alveolar cleft is bone-grafted and all fistulas are surgically closed. Permanent complete palatal retention is necessary, even after alveolar bone grafting. A premaxillary retainer must be placed when the deciduous anterior teeth are lost.

Second Stage. At 7–8 years of age, when the deciduous incisor teeth are replaced by permanent incisors, the anterior crossbite may have to be retreated. The incisors may be rotated and malpositioned. Orthodontic brackets are placed in the upper arch to support a labile arch wire, which will be utilized to reposition the incisor teeth and reduce the premaxillary overbite. A pontic tooth with band is placed on the arch wire to achieve a more pleasing aesthetic result. The premaxilla needs to be properly aligned with the lateral palatal segments prior to alveolar bone grafting; this procedure may be performed between 7 and 9 years of age. A lateral incisor as well as a cuspid may erupt through the area of new bone formation.

Third Stage. Children at 10+ years of age are treated as any other child. The many malocclusion possibilities render it impossible to develop treatment plans for each contingency; instead, basic treatment problems are discussed.

1. **Class I malocclusion (with anterior and/or buccal crossbite):** The crossbite generally is due to lingual bony displacement. The upper arch should be advanced and expanded. If there is sufficient arch length, the missing lateral incisor area is left open. In arch shortage cases, it may be preferable to move the cuspid adjoining the space into the lateral incisor area, rather than extract the first bicuspid on that side. A number of variations of treatment exist according to tooth size and location.

2. **Class II malocclusion:** The anterior overjet may be corrected by retraction of the premaxillary central incisors in bilateral cleft lip and/or cleft palate cases. If one or both lateral incisors are missing, it may be best to place the adjoining cuspid in that space and extract either the opposite lateral incisor or the first bicuspid.
3. **Class III malocclusion:** An anterior crossbite does not necessarily signify that a Class III malocclusion exists. In some cases the anterior teeth can be advanced without excessively increasing their axial inclination. A true depressed midface with poor vertical and anteroposterior development requires midfacial lengthening and advancement. Segmental surgery may have to be utilized to overcome palatal width problems caused by excessive scarring.

7.2.1.3 Use of Orthopedic Forces to Correct Midfacial Recession

Developing anterior crossbites in either mixed or permanent dentition can be corrected using orthopedic protraction forces. These forces must average 800 gm per side and pull downward and forward off hooks placed between the lateral incisor and cuspid. The force needs to be applied 12 h per day. With good cooperation, 5–10 mm of midfacial advancement can be accomplished. The preferred facial mask is the Delaire type.

7.2.2 Supernumerary (Extra) Teeth, Missing Teeth, and Aplasia (Malformed Teeth)

This occurs more frequently in children with cleft lip and/or palate than in other figuration of the nasal floor. (Figs. 7.5, 7.6, 7.13, 7.14). Bishara and coworkers [22] studied untreated adults in India who had clefts of the lip and alveolus only, unilateral cleft lip and palate, and bilateral cleft lip and palate. They observed that the maxilla and cranial base were not different from a matched normal population, but that the relation of the maxilla and mandible to the cranial base varied according to cleft type. Moss [23, 24] Blaine [25], Dahl [26], and Krogman et al. [27] have stated that the cranial base in cleft palate patients differed in both size and shape from noncleft individuals.

The incidence reported in different articles has varied because it is difficult to distinguish between variations rooted in congenital causes and those related to surgery [30]. Recently, it has been observed that supernumerary teeth are more common in the deciduous dentition. Moreover, the incidence of supernu-



Fig. 7.13. The left central and lateral incisor in the line of an alveolar cleft areas reduced in size



Fig. 7.14. Tooth abnormalities. Coalescence of the right deciduous central and lateral incisors in a right unilateral cleft of the lip and alveolus

merary teeth is greatest in cases of cleft lip only and decreases as the extent of the cleft increases. The relationship is the opposite in cases of aplasia; the incidence of aplasia is lowest for cleft lip only and cleft palate only and increases in proportion to the extent or complexity of the cleft [31–35].

In conditions of facial clefting, dental development is, except for the third molars, delayed for all teeth, both maxillary and mandibular [31, 32]. Asymmetrical development of tooth pairs, with delayed development on the cleft side, was recorded in approximately half of a group of children with congenital lip and/or palate clefts [33]. This supports other observations that eruption is delayed in both dentitions [35–54].

Zilberman [55], from a study on clefts of the lip and alveolar structures, and Mirsa and colleagues [56], after investigating clefts of the lip and palate, reported that unilateral clefts are more frequent on the left side and are more common in males than in females.

The incidence of dental malocclusion reported in patients with cleft lip and/or palate varied widely in studies by Huddart and Bodenham [57], Hellquist et al. [58], Dahl et al. [59], Norden and associates [60], Bergland and Sidhu [21], Nylen and coworkers [61], Ranta and colleagues [62], and Hellquist and Skoog [63]. This may be because the patients had varying types of clefts and their cases were recorded at different ages. Rehrman and coauthors [64] found the incidence of malocclusion in the mixed dentition to be twice that in the deciduous dentition.

In cases of cleft palate only, Ranta and colleagues [65] found only a slight increase in anterior crossbite at the transition from the deciduous to the mixed dentition. A noticeable increase in the incidence of anterior crossbite in the mixed dentition, in cases of complete unilateral clefts of the lip and/or palate, was reported by Bergland and Sidhu [21]. This was irrespective of the arch configuration in the deciduous dentition. They also reported that palatal segments stabilized early after lip repair and that further collapse was the exception. However, contrary to the findings just cited, Nylen and coworkers [61] found no increase in the frequency of anterior crossbite in their mixed-dentition group.

7.2.3 Caries

Dahl et al. [66] reported the incidence of caries, gingivitis, and dental abnormalities in preschool children with cleft lip and/or palate in Stockholm, Sweden. Oral health was studied in 49 children 5–6 years old with clefts of the lip and/or palate (CL/P) and 49 healthy children matched for sex and age. The results showed a statistically significant increase in the prevalence and activity of caries among the CL/P children. The average number of decayed and filled tooth surfaces in the cleft group was 7.0 compared with 3.9 in the control group ($p < 0.05$).

The most evident difference between the two groups was found in the number of decayed proximal surfaces. The mean number of decayed proximal surfaces in the CL/P group was 2.5, compared with 0.9 in the control group ($p < 0.001$). No significant differences were found in the prevalence and activity of caries among children with isolated clefts of the lip or palate.

The children with cleft lips/palates also exhibited a significant increase ($p > 0.01$) in the number of gingival units with gingivitis. Other dental abnormalities

included increased enamel hypomineralization ($p < 0.05$), supernumerary teeth ($p < 0.001$), unilateral crossbite ($p < 0.001$), and mesial terminal plane ($p < 0.01$). These results clearly show that children with CL/P as a group must be considered to have an increased risk of caries and gum disease, and should therefore have the benefit of additional preventive programs. (see Chap. 4, Facial and Palatal Growth)

7.3 The Relationship Between the Clefting Process and Contiguous Skeletal Structures

Some studies have indicated that clefting is not an isolated defect but may be a syndrome phenomenon with ramifications in contiguous and often remote structures.

In a study of Danish males, Dahl [26] suggested that the presence of cleft palate, with or without cleft lip, may have ramifications for distant craniofacial structures and their development. Farkas and Lindsay [67] identified consistent variations in facial morphology in the cleft population and concluded that the cleft defect was not an isolated condition. They reported that what might otherwise be considered the normal side of the face in cases of unilateral clefts was not completely normal, and that the anomaly influenced the development of the face equally on both sides.

7.3.1 The Position of the Cleft Maxilla Within the Cranium and the Mandible

Berkowitz [68] undertook a mixed cross-sectional study of CUCLP and CBCLP cases to determine whether the maxillary complex relative to the mandible is posteriorly positioned within the face by studying the dental occlusion. None of the cases had presurgical orthopedics, and the hard palate clefts were closed between 18 and 28 months of age using a modified von Langenbeck procedure with a vomer flap. This study was designed to test McNeil's thesis that the palatal segments, being detached from the nasal septum, are not only reduced in mass but also have not been brought forward with the developing nasal septum. This failure would lead to retrusive midface with a Class III malocclusion.

Berkowitz [68] found that the occlusal relationships at 6 years of age did show Class I and Class II occlusions, but none of the cases had a Class III occlusion, which would have been present if McNeil's [69,

70] hypothesis had been valid. Of the 29 bilateral cases, 5 cases had a crossbite on one side, 1 case had a complete bilateral crossbite, and 6 cases had no crossbites at all. It is quite evident that a buccal crossbite is not, as stated by McNeil, a predictable outcome of the presence of a palatal cleft.

Semb's [71, 72] and Ross's [73, 74] studies and those already acknowledged elsewhere established that in the cleft population both the maxilla and mandible are repositioned within the face (see Chap. 9). However, if McNeil's beliefs were accurate, the bilateral cleft palatal segments would have been left behind in their growth, and a greater proportion of the cases would have shown a Class III malocclusion on one or both sides.

Chierici and associates [75] and Bishara [22, 76] found a relative retrusion of the maxilla and mandible as well as increased steepness of the mandibular plane in various cleft types. Krogman and his colleagues [27] reported significant differences in the cleft population in the size of the cranial base, its configuration, and direction of growth. They concluded that the clefting process has growth and/or development implications for the contiguous cranial base and facial structures as well as for the maxilla.

Bishara [76] reported that the posterior positioning of the maxilla and mandible relative to the anterior cranial base may result from the cleft's influence on contiguous skeletal structures, and that clefting affects maxillary development and facial morphology.

7.3.2 The Cranial Base

Hayashi and colleagues [77] investigated cranial growth of a large sample of subjects from 4 to 18 years of age with complete unilateral clefts. The investigators found that the cranial base angle was flatter, the maxilla was more retruded, and underdevelopment in both the maxilla and the mandible was more pronounced in girls than in boys. They speculated that upper face height in patients of both genders was less than normal as a result of cleft interference with nasal septal and maxillary suture growth and changes in the configuration of the nasal floor. Bishara and coworkers [70] studied untreated adults in India who had clefts of the lip and alveolus only, unilateral cleft lip and palate, and bilateral cleft lip and palate. They observed that the maxilla and cranial base were not different from a matched normal population, but that the relation of the maxilla and mandible to the cranial base varied according to cleft type.

7.3.3 Relationship of the Nasal Cavity to Arch Form (Figs. 7.15, 7.16)

Aduss and Pruzansky [28] wrote that the anatomic distortions common to all of their patients with clefts included marked deviation of the nasal septum toward the noncleft side; flattening, particularly of the inferior turbinate on the cleft side; and an anterolater-

al displacement of the noncleft segment, with an outward and lateral rotation of the premaxillary area adjoining the cleft.

These distortions are the result of unbridled septal growth, deviant maxillary growth, and aberrant vectors of muscle pull. Establishing a continuous muscle band across the cleft, by lip repair, can bring the palatal shelves closer together and modify the config-

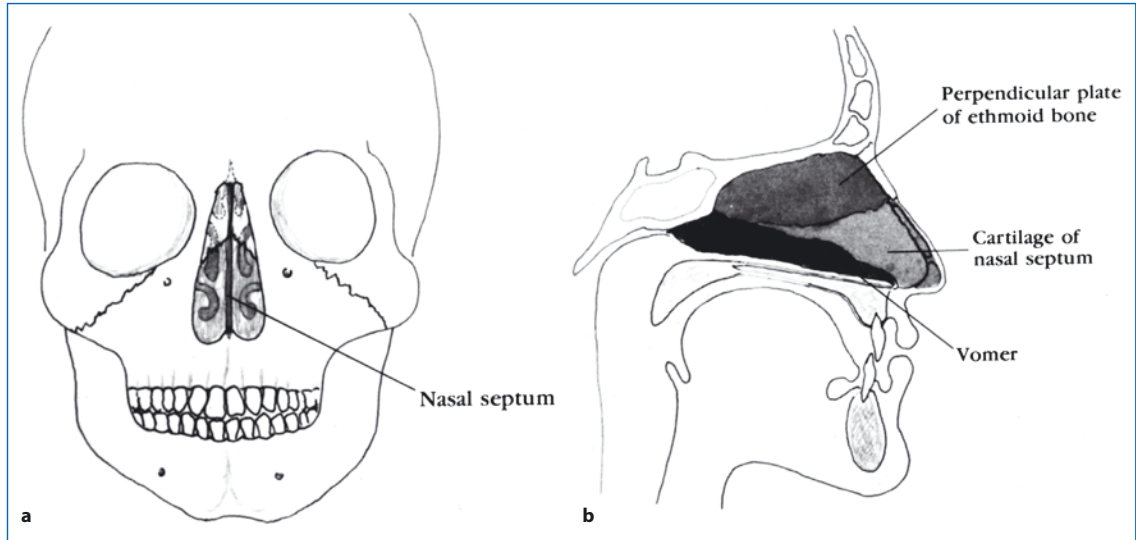
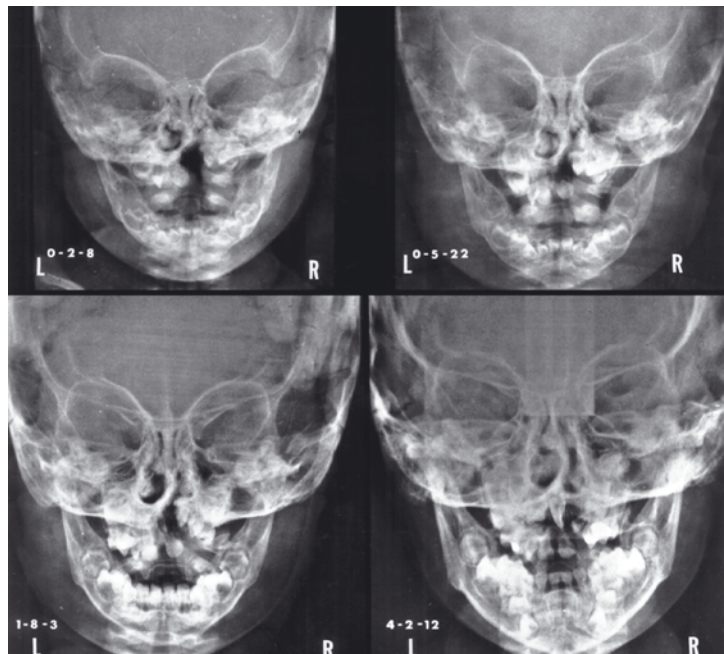


Fig. 7.15. **a** Frontal View. The septum is straight dividing the nasal chamber in two equal parts. **b** Lateral View. The components which make up the nasal septum: the ethmoid bone. The nasal cartilage and the vomer

Fig. 7.16. Serial frontal cephalometric radiographs illustrating geometric changes to the nasal chamber in a CUCLP from birth to 4 years of age before and after palatal expansion. *Top: 0-2-8* At birth a widened nasal chamber is evident. *0-5-22* After lip surgery the nasal chamber has narrowed. *Bottom: 1-8-3* The inferior turbinate on the cleft side (R) makes contact with the vomer. *4-2-12* After palatal expansion the nasal width and the septum to inferior turbinate distance has increased. Because the roof of the mouth is also the floor of the nose, any disarrangement in the architecture of the roof of the mouth is reflected in the nasal chamber. Prior to lip repair, the nasal septum is displaced to the noncleft side. After lip repair with the medial movement of the cleft segment, the septum bows toward the nasal chamber on the cleft side. After palate repair there is continual palatal movement with septal uprighting and decreased septal bowing. The turbinates on the cleft side are flatter, and the buccal teeth on the cleft side may be in crossbite. The nasal chamber on that side is narrowed. (Reprinted from [78])



uration of the palatal segments, as well as the configuration of the internal nares.

Peyton and Ritchie [29], measuring the displacement of the soft tissues of the nose in complete unilateral cleft lip and palate, have shown that deviation of the external part of the nose toward the noncleft side extends for the entire length of the nose, with the greatest displacement at the tip. They further demonstrated that growth of the nasal structures is the same in noncleft children and children with complete unilateral cleft and that the early cleft deformity decreases with time. The natural tendency for self-correction of the septal deviation was evident in the continual uprighting and medial movement of the end of the septum observed in all cases.

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8

Pierre Robin Sequence

SAMUEL PRUZANSKY, JULIUS B. RICHMOND

With some additional photographs by Samuel Berkowitz, DDS, MS

8.1 Growth of Mandible in Infants with Micrognathia [1]

Since Pierre Robin first described the syndrome of hypoplasia of the mandible, cleft palate, glossoptosis, inspiratory retraction of the sternum, cyanosis, and malnutrition which has come to bear his name [2, 3], numerous reports of cases have appeared in the literature. The fatal termination of some cases is a testimony of the serious nature of the syndrome. Although a variety of mechanical and surgical therapeutic procedures have been suggested [4–11], a rationale for the proper management of individual patients has not been adequately presented. It is the purpose of this communication to present observations from the serial studies of patients with this disorder, upon which a physiologic approach to management may be based.

It is generally agreed that the pathophysiologic events in this syndrome are as follows: the receding chin fails to support the tongue in its normal forward relationship and hence fosters the glossoptosis. The retroposed tongue impinges against the posterior wall of the pharynx, obstructing inspiration and impeding feeding. Slight excess in mucus or saliva tend to enhance the pharyngeal obstruction and may precipitate severe cyanotic seizures, resulting in death. Starvation and respiratory infections, or both, may follow as a consequence of the chronic glossoptosis.

Since the major symptoms are definitely related to the micrognathia, it was of special interest to focus our attention upon the growth of the mandible in these infants. In reviewing the published case reports, it appeared that two questions, basic to an understanding of a more appropriate management of these cases, remained unanswered. The first question could be phrased as follows: How soon would mandibular growth be sufficient to accommodate the tongue and hence insure a more adequate airway? This is assum-

ing that the infant could be placed in an adequate metabolic climate. The answer to this question would have an important bearing on immediate management. The second question is related to whether or not mandibular growth would be sufficiently sustained to provide an esthetically satisfactory facial profile.

The answer to these questions had to await the development of accurate roentgenographic techniques for measuring the growth of the head in infants, which made it possible to undertake a serial study of the growth of the micrognathic mandible. The results of these investigations have provided useful information as a basis for therapy and prognosis.

During the past 25 years, cephalometric roentgenography has produced valuable information pertaining to the growth of the head in normal [12] and pathologic conditions [13] in man. In addition to the cephalometric roentgenographic measurements, dental impressions of the maxillary arch were obtained at regular intervals. The impressions were cast in dental stone and then subjected to various measurements. The infants were recalled for study every 3 months during the first year of life and twice annually until the age of 5 years. Thereafter, they were observed annually.

From a large series of similar cases now under longitudinal study at the Cleft Palate Center of the University of Illinois [14] we have selected three cases for presentation. At birth, each of these infants present an isolated cleft of the palate and mandibular micrognathia. Despite these similarities, certain important differences existed to vary the management and progress of each infant. The differences to be described in each of these cases represent the major variations which we have encountered in our experience with this syndrome. For the sake of brevity, the case history will be confined to such information as is directly pertinent to the purpose of this paper.

8.1.1 Case 1

At 2 months of age, D.R.P., a small white dehydrated baby girl, was admitted to the Research and Educational Hospital, with the principal complaint of intermittent pneumonitis for the previous 5 weeks. After a normal full-term spontaneous delivery, it was noted that the baby had an isolated cleft palate and a small mandible. The birth weight was 5 lb. 13 oz. (2,640 g). Feeding had been difficult because of the tendency of the baby's tongue to fall back into the pharynx. There was no history of a cleft on either side of the family. The mother gave no history of being exposed to a contagious disease or other illness during her first trimester of pregnancy.

Because of the patient's congenital defects, she presented primarily a feeding problem. An attempt was made to design an obturator so that the baby could be bottle-fed. This was met with failure, and gastric gavage was necessary. However the patient did not gain weight and continued to do poorly. Approximately 1 month after admission, it was considered that a tracheotomy was the only procedure which might possibly save the baby's life. After the tracheotomy, respiration was greatly facilitated, and soon the patient was removed from the oxygen tent. Her improvement thereafter was constant. The patient was discharged from the hospital 3 months after admission, at 5 months of age.

Growth Studies: Oral examination disclosed a cleft involving the one half of the hard palate and extending posteriorly throughout the length of the soft palate. Ptosis of the tongue was evident.

On the fifth day following the tracheotomy, at the age of 3 months 1 day, the first lateral film was obtained without sedation. A tracing of this film revealed that the head was in slight dorsiflexion, the position of greatest comfort for the infant. The relative smallness of the mandible and its effect on the facial profile was self-evident. In relation to contiguous anatomical structures, the posture of the tongue was abnormal. It projected through the cleft in the palate into the nasal cavity. (Fig. 8.2)

In the lateral head plate, the dorsum of the tongue was visible at a level above the palatal plane. Posteriorly, the tongue impinged on the airway. The posterior surface of the tongue, just above the epiglottis, was in close approximation to the posterior outline of the pharyngeal wall. At this level, the airway was almost completely occluded. The tracheotomy tube was in place. The airway in its posteroanterior dimensions is fairly wide, and the tongue occupies a more protrusive relationship to the mandible.

The second film in Fig. 8.2, obtained less than 6 weeks later, at the age of 4 months 13 days, revealed considerable enlargement of the airway. At the same time, a considerable increment in the growth of the head and, above all, of the mandible was recorded. Subsequent films, up to the age of 3 years 5 months 12 days, gave proof of the gradual and continued growth of the lower jaw in relation to the total face and the increase in the dimensions of the pharyngeal airway. At 8 months following removal of the tracheotomy tube, the air passage was demonstrated to be quite adequate (Fig. 8.2).

The superimposed tracings, from 3 months 1 day of age to 3 years 5 months 12 days, reveal the progressive

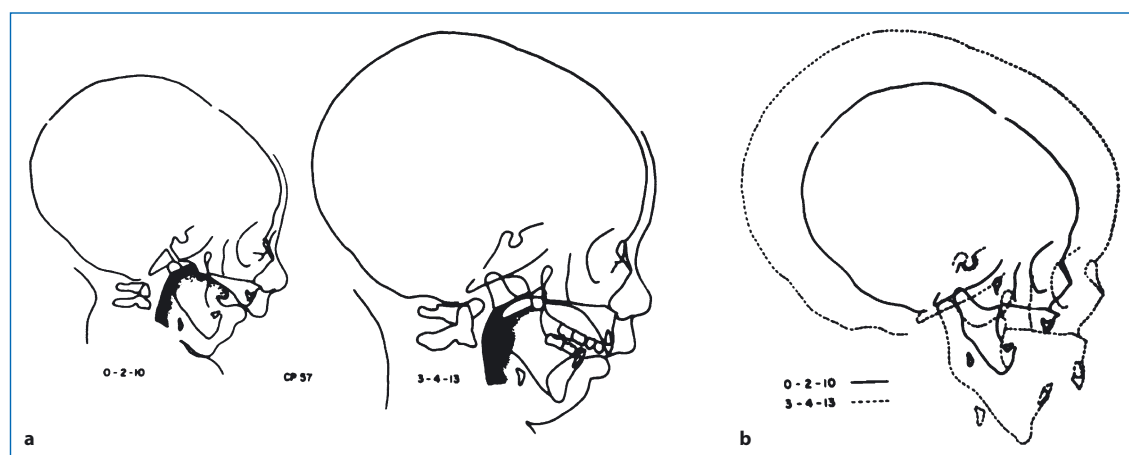


Fig. 8.1. **a** Serial cephalometric tracing of a child with Pierre Robin Sequence showing a severely micrognathic mandible. At 3-4-13 there is a big increase in the pharyngeal space (darkened area). **b** Superimposed tracings at 2 months and 10 days

(0-2-10) and 3 years, 4 months, and 13 days (3-4-13) shows a rapidly growing micrognathic mandible. (Reprinted with permission from [1])

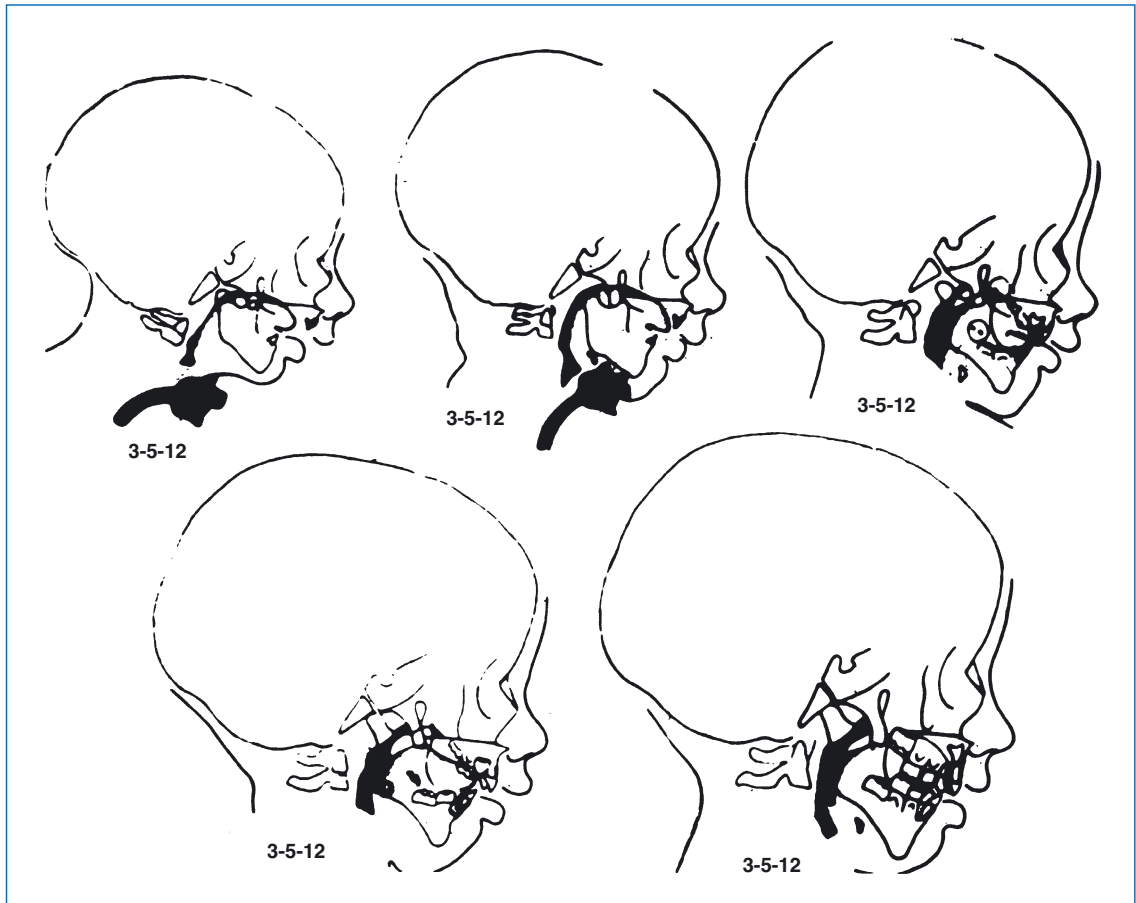


Fig. 8.2. Pierre Robin sequence. A series of tracings of the lateral film from 3 months, 1 day of age (0-3-1) to 3 years, 5 months, 12 days (3-5-12). The pharyngeal airway is filled in (black), and the stippled area denotes the border of the tongue. A tracheotomy tube is visible in the first two films. Soon after birth (3 and 4 months of age) the dorsum of the tongue is visible at a level above the palatal plane within the palatal cleft space. Posteriorly, the tongue just above the epiglottis impinging on the airway.

At this level, the airway was almost completely occluded. Comparison with the remaining three figures, ages 0-8-10, 1-1-0 and 3-5-12, reveals the configuration of these structures under normal circumstances after closure of the cleft. The airway in its posteroanterior dimensions is fairly wide, and the tongue occupies a more protrusive relationship to the mandible. (Reprinted with permission from [1])

growth of the cranial vault, the maxilla, and the middle face and the increase in length and height of the mandible (Fig. 8.1b). Gradual improvement in the facial profile was recorded by the changes in the facial angle and in the angle of convexity. The facial angle is a measure of the degree of protrusion or recession of the chin. In this instance, the facial angle increased from 64°, at 3 months of age, to 70°, at the age of 3 1/2 years, indicating a reduction by 6° in the recessiveness of the chin. While the mandible is still in a retrusive relation to the rest of the face, the potential for further improvement with continued growth still exists.

The changes in the angle of convexity were more interesting. This measurement relates the maxilla to

the total facial profile. At 3 months of age, the angle of convexity was 140°, and at 3 1/2 years it measured at 154°. The integrated growth of the several areas of the face was such as to improve the overall configuration of the facial profile. Serial photographs at 2 months, at 13 months, and at 3 years of age further testify to the changes in this child's face. In the last photograph, the patient is posed beside her older sibling.

The changes in the position of the hyoid bone are of particular interest, insofar as they reflect a change in the relative position of the tongue. The tongue is composed of several individual muscles originating from the base of the skull, the mandible, the hyoid bone, and the walls of the pharynx. Changes in the position of any of its bony or fibrous attachments would

tend to reflect on the position of the tongue. Conversely, changes in the posture of the tongue would reflect on the spatial relations of the mandible and hyoid bone. Therefore, to study the position of the hyoid bone is, in a sense, to study the position of the tongue. With growth there occurs a forward and downward migration of the hyoid bone from the base of the skull. The pattern of changes in the posture of the hyoid bone observed in this patient sheds further light on the favorable adjustments consequent to growth. During the first 5 months of our studies, the hyoid bone migrated downward and forward. This resulted in an increase in the angle S-N-H. But, from 8 months onward, this angle became fairly stable and the hyoid bone began to descend principally in a downward direction.

Comment: This case was selected to typify the findings in several similar cases, one of which has been followed to the age of 7 years. Not all cases of Pierre Robin syndrome present such acute histories. When clinical evaluation suggests that there will be no improvement or that possibly death may ensue, tracheotomy should be undertaken without hesitation to prevent further aggravation of the symptoms. Once an adequate respiratory exchange was made possible, improvement in oxygenation and feeding followed. In such instances, we have recorded rapid growth and favorable changes in the facial appearance.

8.1.2 Case 2

J.G, a white girl, was referred to the outpatient clinic of the Cleft Palate Center at the age of 2 months with a diagnosis of cleft palate and mandibular micrognathia. Following an uneventful pregnancy, the delivery was normal and at full term. The birth weight was 6 lb. 11 oz. (3,030 gm). The infant had some difficulty in breathing, but this was relieved by placing her in a prone position. Tube feeding was employed for the first few days after which she was given bottle feedings. At 6 days of age, the infant was discharged from the hospital. There was no family history of cleft palate. The mother suffered no illness during her pregnancy.

Oral examination revealed an unusually small tongue closely attached to the floor of the mouth. In the course of our first examination under sedation, the infant became cyanotic and failed to initiate mandibular movements sufficient to permit the passage of air. This was relieved immediately by maintaining forward traction on the tongue and mandible. After about 5 min, the infant recovered control of mandibular movements, and respiration normally. Aside from this isolated episode, which occurred un-

der sedation, the parents did not report any similar difficulties. The child has continued to grow and develop at a satisfactory rate.

Growth Studies: The casts disclose symmetrical cleft of the hard and soft palate, extending distally from the region of the nasopalatine foramen. Additional casts obtained at regular intervals revealed that the cleft had narrowed, so that it now presents a narrow V-shaped defect (Figs. 8.3, 8.4).

The earliest lateral head palate, at 2 months 10 days of age, displayed a small mandible and small tongue. The latter was positioned high and above the floor of the nose, but relatively remote from the posterior wall of the pharynx. The airway appeared sufficient to sustain respiration without any undue effort on the part of the infant. Progressive growth changes recorded up to the age of 3 years, 4 months, 13 days disclosed mandibular growth and generalized growth in all areas of the face and cranial vault. Mandibular growth was continuous and progressively downward and forward. During the period studied, from 2 to 40 months of age, the facial angle increased from 61.5, becoming more obtuse. The angle of convexity increased from 147° to 155°. Altogether, the changes were in a direction tending to minimize the recessiveness of the chin in relation to the rest of the face.

Comment: Micrognathia by itself is not sufficient to produce glossoptosis and respiratory embarrassment. If the tongue is large or even normal in size, the small recessive mandible will tend to displace the tongue distally and superiorly. It is this displacement that produces the respiratory obstruction both into the hypopharynx and into the posterior choanae. On the other hand, if the tongue is small, there will be no obstruction of the airway even in the presence of a micrognathic mandible. In this instance, the simultaneous occurrence of micrognathia and microglossia averted the respiratory difficulties commonly experienced in such instances.

The tendency to lose reflex control of the muscles of respiration and deglutition under anesthesia or sedation renders such procedures unusually hazardous in these patients because of the limited reserve. It is, therefore, important that such procedures be undertaken with full knowledge and anticipation of possible respiratory obstruction, in order that adequate emergency provisions for the establishment of an airway be available.

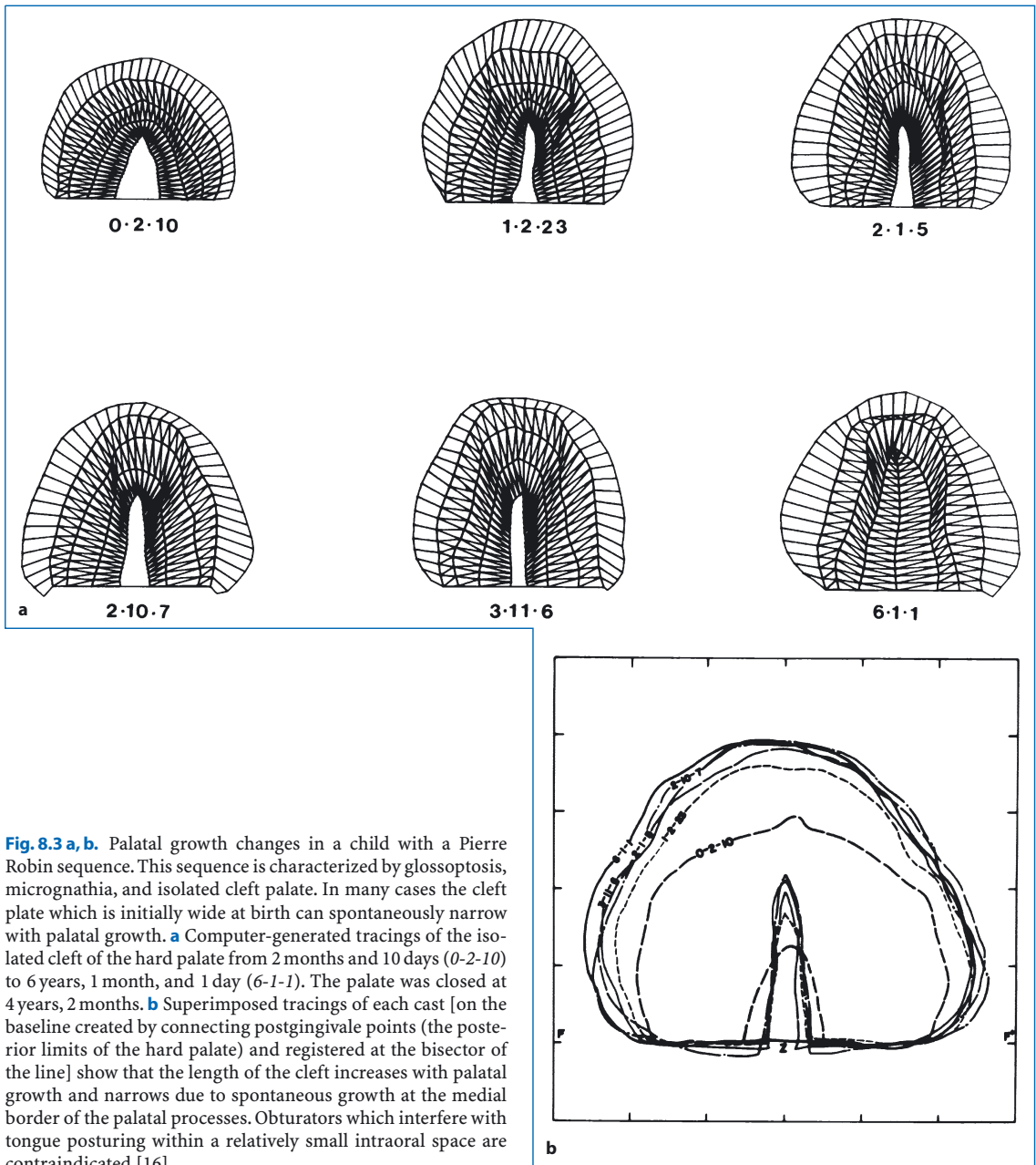


Fig. 8.3 a, b. Palatal growth changes in a child with a Pierre Robin sequence. This sequence is characterized by glossoptosis, micrognathia, and isolated cleft palate. In many cases the cleft plate which is initially wide at birth can spontaneously narrow with palatal growth. **a** Computer-generated tracings of the isolated cleft of the hard palate from 2 months and 10 days (0-2-10) to 6 years, 1 month, and 1 day (6-1-1). The palate was closed at 4 years, 2 months. **b** Superimposed tracings of each cast [on the baseline created by connecting postgingivale points (the posterior limits of the hard palate) and registered at the bisector of the line] show that the length of the cleft increases with palatal growth and narrows due to spontaneous growth at the medial border of the palatal processes. Obturators which interfere with tongue posturing within a relatively small intraoral space are contraindicated [16]

8.1.3 Case 3

E.C., a white boy aged 5 weeks, was referred to the outpatient clinic of the Cleft Palate Center for longitudinal growth studies. The delivery had been normal and at full term. His birth weight was 7lb., 8oz. (3,400 gm.). There was no history of cleft on either side of the family. No difficulty in breathing was encountered, and the infant was discharged from the

hospital on the sixth day. After a brief adjustment period, the infant was readily fed by a combination of a hard nipple and by means of a premature baby bottle nipple.

Some snoring sounds were heard, especially as the infant was placed on its back and the head elevated with slight ventroflexion on the chest. The infant preferred to sleep on either side, and in these positions the snoring sounds were at a minimum. This baby

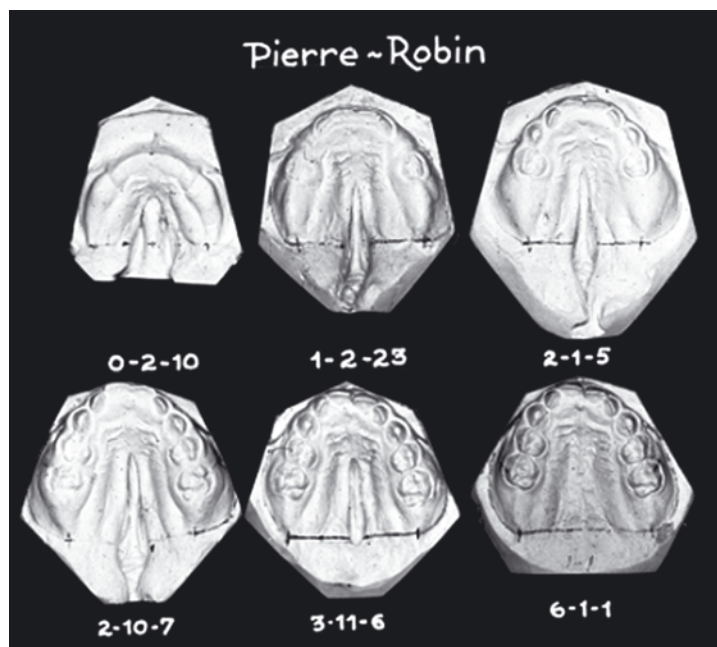


Fig. 8.4. A series of cast of the maxillary arch from 2 months 10 days of age to 6 years 1 month. Note the progressive narrowing in the lateral dimension of the cleft. (Reprinted with permission from [1])

showed progressive improvement, and at the age of 5 months he weighed 15 lb., 8 oz. (7,030 gm.)

Growth Studies: Two sets of records are available in this case. The first was obtained at the age of 1 month 7 days, and the second at 3 months 25 days of age.

The first cast of maxilla revealed a wide parabolic cleft extending distally from the nasopalatine foramen. The widest portion of the cleft, at the level of the maxillary tuberosities, measured 16 mm. Although the second cast exhibited an increase in the length and width of the palate, there was a decrease of 1.5 mm in the width of the cleft at its widest portion. During the first examination, the tongue was observed to occupy at rest the opening into the nasal chambers provided by the cleft in the palate. This was further confirmed by examining the frontal and lateral views of the head plates. The second series of films indicated that the tongue was now postured in a more inferior position and no longer occupied the nasal cavity to the same extent previously noted. This new position of the tongue could be explained by the downward and forward growth of the mandible that had occurred in the interim.

In the first lateral film, the recessive chin, the distally and superiorly malposed tongue, and relatively restricted airway were clearly observed. Two and one-half months later, considerable growth in the mandible had occurred to improve the facial profile, alter the posture of the tongue, and increase the anteroposterior diameter of the airway. The tongue was

no longer in close apposition to the posterior pharyngeal wall, and its superior margin did not extend into the nasal cavity to the degree previously observed. Coincidentally, the mother reported a diminution of the stertorous breathing that had been present.

Superimposition of the tracings of the bony structures revealed the rapid growth characteristic of this early period in life. In 2 1/2 months, that cranial vault and all parts of the face exhibited proportionate increases. Particularly encouraging was the amount and direction of growth displayed by the lower jaw. Mandibular growth was responsible not only for reducing the glossoptosis and increasing the airway, but for the improvement in the appearance of this baby's face.

Comment: The problem presented by this baby was unique and different from the two previous cases of the partial obstruction of the airway. Diligent nursing care to determine the most comfortable position for breathing and feeding may be sufficient to tide such cases through their critical period. In some instances the prone positioning and orthostatic feeding suggested in the literature are most successful. Again, one is impressed by the remarkable potential for prolific growth during this period of life; a potential that is shared by the small mandible. It follows then, that every effort must be made to permit the realization of the baby's potential for growth by providing an adequate airway, which, in turn, facilitates feeding. The clinical course to be followed is varied and depends on

the severity of the symptoms and principally upon the degree of obstruction of the airway.

8.2 Comment

The representative sampling of cases presented provides an answer to the questions which the study was designed to solve. It is observed from the data presented that the mandible possesses remarkable potentialities for growth in patients with the Pierre Robin syndrome. Thus, all efforts should be directed toward sustaining life in a metabolically favorable climate in order that a more physiologic airway may be established as growth proceeds. With growth, the glossoptosis is minimized and spontaneous resolution of the respiratory and feeding problems occurs. It is our opinion that tracheotomy should be resorted to promptly if respiratory embarrassment is significant, in order to achieve a sufficient airway to provide adequate oxygenation. This is undoubtedly a life-saving procedure in some patients.

On the basis of our longitudinal growth studies, certain prognostications concerning the future growth of the micrognathic mandible are permissible. In most instances, the increment in mandibular growth, as related to total facial growth, is sufficient to overcome the extreme recessiveness of the chin that is observed at birth. Since mandibular growth continues until late adolescence, it is possible to hope for an esthetically pleasing profile in adulthood. The management of the cleft palate has been in keeping with the established criteria for the treatment of palatal defects.

The lateral cephalometric film served as a valuable diagnostic tool in estimating the degree of obstruction of the airway as a result of the glossoptosis. In our experience, there was a high positive correlation between the degree of obstruction revealed in the x-ray film and the incidence and severity of the respiratory difficulties. When obstruction of the air passage was complete and the tongue was practically in contact with the posterior wall of the pharynx, tracheotomy was recommended as a life saving procedure. If the obstruction was incomplete, more conservative measures were employed. Care was taken to ascertain the most comfortable postures for breathing and feeding for the individual case, and the nurse or parent was carefully instructed in the care of the infant. Appropriate nipples were selected to minimize the energy expended by the infant in the feeding process.

In the course of these studies, we were aware of an obvious objection to placing so much reliance on these roentgenograms. Since many of these films were obtained under mild sedation, was it not possible that the posture of the mandible or of the tongue might

have been altered by the sedative? Secondly, the film depicted a static view of the airway and represented only two dimensions. Did this view properly reflect the kinetic ability of the infant to manipulate the tongue and jaw hence the consistent correlation between the findings in our films and the clinical state? Moreover, when the films were repeated in the same infant without sedation, similar postures were recorded for the structures under analysis. It was important that the postures of the head in relation to the neck be kept constant. Dorsiflexion or ventroflexion of the head varied to posture of the mandible and tongue and produced changes in the configuration of the airway. To indicate alterations in the posture of the head to the neck, our tracings purposely included at least the first two cervical vertebrae.

We recognize that few institutions possess cephalometric roentgenographic equipment. Therefore, we should like to point out that an ordinary lateral film obtained by carefully positioning the infant can provide useful diagnostic data. To minimize enlargement, a target-object distance of at least 3 ft. (90 cm) is recommended. For the sake of definition and to further decrease enlargement, the object-film distance should be kept at a minimum. Sjölin [15] has published interesting films to describe his experiences with a case of micrognathia. Although his films did not permit quantification of the growth changes, they were adequate for diagnostic purposes.

A number of papers in the literature claim to "stimulate" the growth of the mandible by a variety of mechanical devices or surgical procedures. For example, a special nursing bottle was designed to force the infant to protrude his jaw in order to obtain nourishment and, by this protrusion, to stimulate mandibular growth [4]. From our data, we would conclude that the nursing care enabled the infant to survive until mandibular growth was sufficient to provide a more adequate airway.

In another report, continuous traction on the mandible was maintained by circumferential wiring around the symphysis. The authors claimed growth-stimulating properties for this procedure [10]. From the findings in our series, it would seem that mandibular growth probably occurred spontaneously and not because of the stimulus provided by surgical traction.

The important and prime objective in the care of these children is to provide an airway. If possible, this should be accomplished with a minimum of trauma. Secondly, the infant's total needs should be assessed to provide optimal conditions for somatic growth. As the potential for growth is permitted to express itself, the chin grows downward and forward away from the base of the skull. With this pattern of growth, adequate space for the tongue is provided, the airway

enlarges, and there follows a spontaneous resolution of the symptoms. Also, there are progressive improvements in the facial appearance.

There is another dimension to the abnormal posture of the tongue, as observed in these patients, that merits discussion. Not only does the tongue block the pharyngeal processes and hence prevent their fusion. The high incidence of micrognathia in the population of clefts involving only the hard and soft palate lends support to this theory. Mandibular micrognathia is a physiological finding in early intrauterine life. If for some reason the micrognathia persists and fails to carry the tongue down and out of the nasal cavity, a cleft in the palate might result.

In early postnatal life, the tongue acts to keep the cleft palatal processes apart. As the tongue descends with mandibular growth and no longer forcefully intrudes itself into the nasal cavity, the palatal processes tend to approximate in the midline. Fusion of the palatal processes cannot occur, but the narrowing in the clefts is recorded fact.

8.3 Summary and Conclusions

The development of the accurate techniques for cephalometric roentgenography of infants has made possible a longitudinal study of the growth of the micrognathic mandible. As a result of these studies, useful diagnostic and prognostic information has been obtained to provide a rationale for the management of individual cases.

The lateral cephalometric roentgenogram is a valuable diagnostic aid in assessing the severity of the glossoptosis and its obstruction of the airway. A definite correlation exists between the degree of constriction of the airway and the severity of the clinical state. On the basis of these findings, it is possible to recommend either conservative management or tracheotomy in extreme situations, or distraction osteogenesis. Three cases, out of a larger series of similar cases, were presented to indicate the spectrum of variations to be encountered.

In all instances, it was found that where an adequate metabolic situation was provided and the infant gained weight, mandibular growth during the first few months was sufficient to provide for a natural resolution of the symptoms attending the glossoptosis.

Longitudinal records have indicated that mandibular growth is proportionally adequate to reduce the

retrognathic profile and provide an esthetically harmonious facial appearance.

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9

Characteristics of Facial Morphology and Growth in Infants with Clefts

SVEN KREIBORG, NUNO V. HERMANN, TRON A. DARVANN

9.1 Introduction

Congenital clefts of the lip and/or palate can arise in isolation or together with other malformations (syndromes) [28]. This chapter deals solely with “nonsyndromic” clefts.

Both individuals with unoperated and operated clefts have a face which differs from those of unaffected individuals. Since the introduction of roentgen-cephalometry more than 70 years ago [8] hundreds of cephalometric studies, including both unoperated and operated cleft individuals have suggested that some deviations are directly caused by the primary anomaly, while others are caused by the surgical interventions and the following dysplastic and compensatory growth of the facial bones [e.g., 1, 3, 4–10, 12, 15, 18–27, 29, 30, 48, 50, 51, 53–55, 57, 59–61]. However, the relative importance of the intrinsic factors, the iatrogenic factors, and the functional or adaptive factors for the facial development is still unclear. There are probably several reasons for this. Firstly, comprehensive knowledge of craniofacial morphogenesis in cleft newborns or infants before surgery, based on large, consecutive, well-controlled samples, is very scarce. This situation is not surprising since, in developed countries, the cleft of the lip is surgically treated within the first couple of months after birth. Thus, the possible period of examining the unoperated state is short and several methodological problems are involved. Secondly, the cephalometric analyses are most often limited to the lateral projection using simplistic cephalometric analyses, typically based on 15–20 reference points, and almost invariably measuring maxillary prognathism as the *S-N-A* angle or similar measurements to the premaxilla, and the use of infant cephalometry has been very limited. These authors are of the opinion that incomplete knowledge about the intrinsic factors related to the cleft anomaly has automatically lead to excessive emphasis on the importance of iatrogenic and adaptive factors in facial development of cleft children.

9.2 The Danish Experience

In the middle of the 1970s we decided to take advantage of the very favorable sampling conditions in Denmark in an effort to contribute to the question of the characteristics of facial growth and development in children born with clefts [42]. In Denmark, for more than 65 years, all newborns with facial clefts have been recorded at the Institutes for Speech Disorders in Copenhagen and Århus. Repeated follow-up examinations have shown that the registration of clefts in Denmark is highly reliable and nearly complete. The population is homogeneous and stable, and only very few children are lost to follow-up. Furthermore, all primary cleft surgery is performed in one hospital by one surgeon.

Inspired by [52] we constructed a three-projection infant cephalometer, which can obtain truly orthogonal lateral, frontal, and axial cephalograms [43]. A comprehensive cephalometric analysis system was developed including all craniofacial regions (calvaria, cranial base, orbits, maxilla, mandible, airway, cervical spine, and soft-tissue profile) [44, 32, 36], and the method was validated [36]. Furthermore, new methods of visualization of differences in craniofacial morphology and growth between different groups were developed using mean plots [36, 44], color-coded vectorgrams [36], and color-coded surfaces on a 3D CT-model [16].

During the 6 years from 1976 to 1981, there were 359,027 live births in Denmark. A total of 678 newborns of Northern European ancestry with cleft lip, cleft palate, or both were registered in the period. Twenty-four infants died before 22 months of age, and for practical reasons material uptake had to be omitted in some patients with isolated cleft palate. Only nonsyndromic clefts were included in the study, but 602 of the 678 children (about 90%) were examined by us [42] and nearly all at both 2 months of age (before any surgical or orthopedic treatment) and at 22 months of age (before closure of the posterior

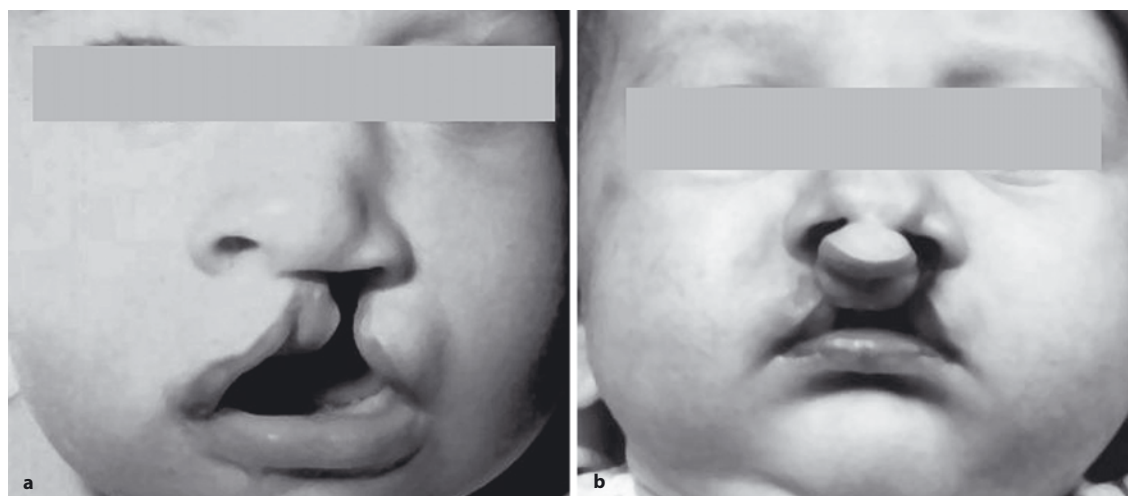


Fig. 9.1. **a** The facial morphology in a 2-month-old unoperated infant with unilateral complete cleft lip and palate (UCCLP). **b** The facial morphology in a 2-month-old unoperated infant with bilateral complete cleft lip and palate (BCCLP)

Table 9.1. Summary and comparison of the most important findings in the primary anomaly in children with RS, ICP, BCCLP, and UCCLP*

Anomaly	RS	ICP	BCCLP	UCCLP
Maxilla				
Decreased length measured to premaxilla (<i>sp-pm</i>) ^a	+ ¹	+	− ²	−
Retrognathia measured to premaxilla (<i>s-n-ss</i>) ^b	+	+	− ³	−
Decreased posterior length (<i>ci-pm</i>) ^c	+	+	+	+
Retrognathia measured to base of jaw (<i>s-n-ci</i>) ^d	+	+	+	+
Decreased posterior height	+	+	+	+
Increased width	+	+	++	++
Nasal cavity				
Increased width	+	+	++	++
Mandible				
Decreased length	+++	++	++	+
Retrognathia	+++	++	++	+
Pharyngeal airway				
Reduced size	+++	++	+	+

* RS, Robin Sequence; ICP, Isolated Cleft Palate; BCCLP, Bilateral Complete Cleft Lip and Palate; UCCLP, Unilateral Complete Cleft Lip and Palate.

¹ The deviation from the norm is shown as + or −, meaning, e.g., that decreased total length of the maxilla was observed in the ICP and RS groups but not in the UCCLP group, and that the length of the mandible is decreased in the UCCLP group, very decreased in the ICP and BCCLP groups, and severely decreased in the RS group.

² The total length was significantly increased.

³ The prognathism was increased measured to the premaxilla.

^a *sp-pm*: Anterior nasal spine to point *pterygomaxillare*.

^b *s-n-ss*: S-N-A.

^c *ci-pm*: Point *crista infrazygomatrica* to point *pterygomaxillare*.

^d *s-n-ci*: Maxillary prognathism measured to the infrazygomatic crest.

palate in the children with clefts of the secondary palate). All children were treated by the same surgeon (Dr. Poul Fogh-Andersen), and in the children with cleft of the primary palate the cleft lip was, in all cases, closed using a Tennison procedure. One third of

the children had isolated cleft lip (CL), about 40% had combined cleft lip and palate (CLP), and about 27% had isolated cleft palate (CP). The clefts were subclassified according to the method of Jensen et al. [42].

In the 602 children included in the study, cephalograms were obtained in the lateral, frontal, and axial projections by three experienced orthodontists (Dr. Birgit Leth Jensen, Dr. Erik Dahl, and Dr. Sven Kreiborg). In addition, impressions were made of the maxilla, and anthropometric registrations (body height, body length, and head circumference) were carried out. The results of the cephalometric analyses have been presented in a number of publications [13, 14, 17, 33–41, 45–47]. So far, we have analyzed infant craniofacial morphology and early craniofacial growth in detail in three dimensions in the following groups: incomplete unilateral cleft lip (UICL), isolated cleft palate (ICP), Robin sequence (RS), unilateral complete cleft lip and palate (UCCLP) (Fig. 9.1a), and bilateral complete cleft lip and palate (BCCLP) (Fig. 9.1b). In the following, we shall summarize our findings, with emphasis on the unoperated infant to shed light on the intrinsic factors related to the cleft condition (see Fig. 9.2 and Table 9.1), and compare them to data in the literature on unoperated adolescents and adults with clefts.

9.2.1 Cleft Lip (CL)

Isolated CL involves only structures of the embryonic primary palate. The craniofacial morphology in CL subjects has been shown to be fairly normal except for the small region of the cleft including the premaxilla and the incisors. In unoperated bilateral complete CL the premaxilla may, however, protrude markedly. In unilateral, *complete* CL the protrusion is less pronounced but asymmetric. In subjects with unoperated, unilateral, *incomplete* cleft lip UICL the protrusion of the premaxilla is negligible [33]. The interorbital distance in CL subjects seems to be slightly increased compared to the norm (11). The basal part of the maxilla has a normal prognathism in relation to the anterior cranial base, and the mandible is of normal size, shape, and inclination [12; 33]. Following lip surgery, the premaxilla is molded into a normal position, and maxillary prognathism measured to the *point A* or *ss (subspinale)* is normal [12, 31, 33–35]. In conclusion, subjects with UICL have a very close to normal craniofacial morphology from infancy to adult age, and consequently, we have used our group of infants with UICL as a control group in the study of deviations in craniofacial morphology and growth of infants and young children with ICP, RS, UCCLP, and BCCLP, since no actual normative cephalometric data for Danish infants and young children are available.

9.2.2 Cleft Palate (CP)

Isolated cleft palate (ICP) involves only structures of the embryonic secondary palate. In Fig. 9.2a, the mean facial diagrams of the ICP group is superimposed on the mean facial diagram of a group of age-matched infants with UICL (control group). The major deviations in the ICP group were: reduced length and posterior height of the maxilla; maxillary retrognathia; increased width of the maxilla and the nasal cavity; and reduced length of the mandible with mandibular retrognathia. Thus, the ICP group revealed *bimaxillary* retrognathia. The sagittal jaw relationship was, however, normal. In addition, in the ICP group the upper airway dimensions were reduced.

Bimaxillary retrognathia and a short mandible were previously documented in unoperated older children [60] and adults with ICP [12, 2].

9.2.3 Robin Sequence (RS)

Robin sequence (RS) is defined as a triad of symptoms: isolated cleft palate, micrognathia, and glossop-tosis [28]. RS may be part of several syndromes, e.g., Treacher-Collins syndrome [11, 46]. In this chapter, only nonsyndromic cases of RS will be discussed. We consider this group as a subgroup of the ICP group [39]. In Fig. 9.2b, the mean facial diagram of the RS group at 2 months of age is superimposed on the mean facial diagram of the control group. The major deviations in the RS group were: decreased length and posterior height of the maxilla; maxillary retrognathia; increased width of the maxilla and nasal cavity; very short mandible with marked mandibular retrognathia. Thus, the RS group revealed *bimaxillary retrognathia*; the retrognathia was, however, most marked for the mandible and the sagittal jaw relation was increased. In addition, the RS group had a significantly smaller cranial base angle (*n-s-ba*) resulting in a smaller depth of the bony nasopharynx than the controls, and the upper airway dimensions were markedly reduced. The degree of maxillary retrognathia was similar in the RS and the ICP group. However, the mandibular retrognathia in the RS group was even more marked than in the ICP subjects. It would seem that RS subjects probably represent the extreme part of the ICP population in terms of mandibular retrognathia and upper airway constriction. As mentioned above, we consider the RS group as a special subgroup of the ICP group. Accordingly, we believe the bimaxillary retrognathia to be intrinsically associated with the cleft of the secondary palate.

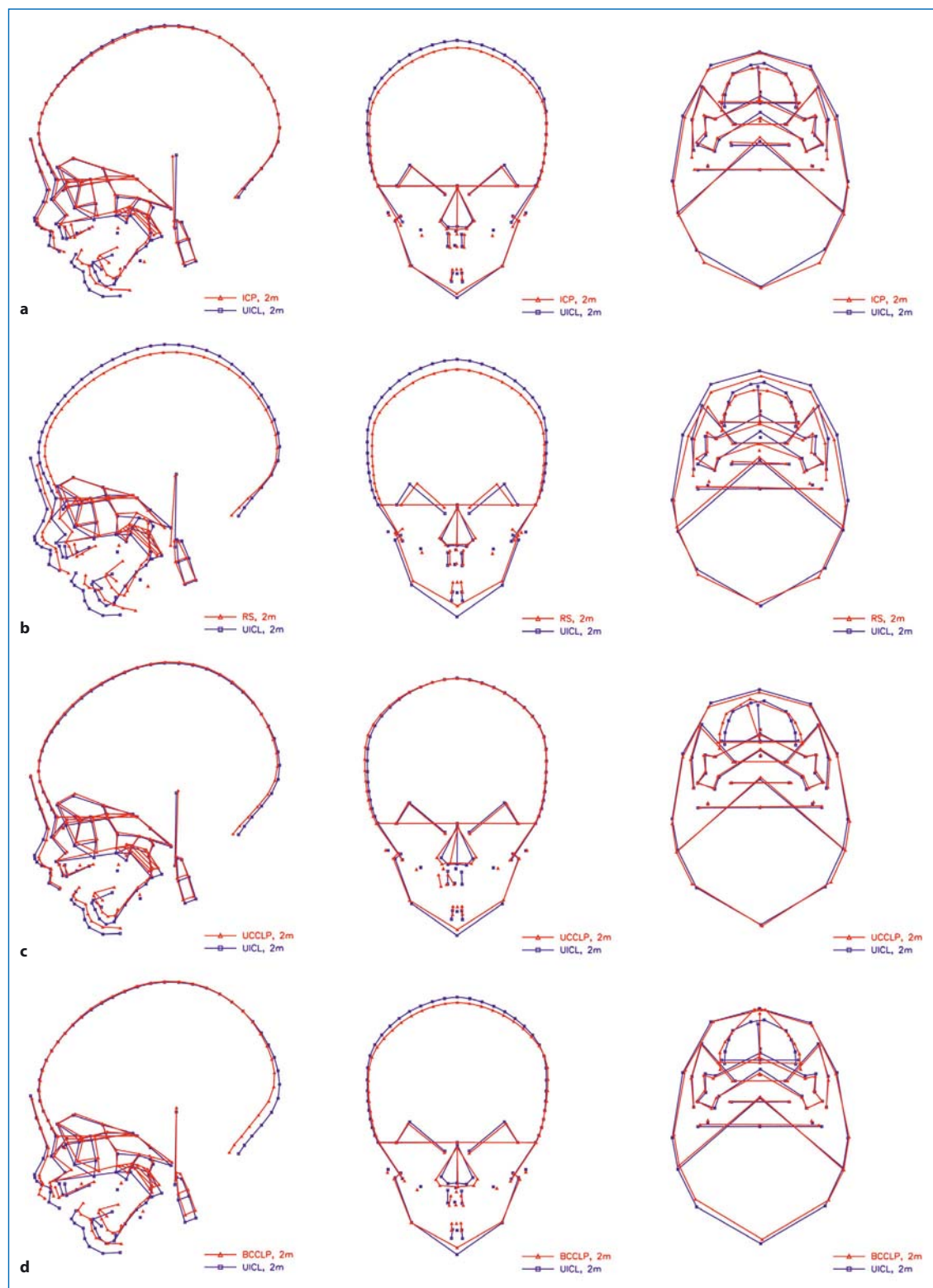


Fig. 9.2 a–d. Mean plots in three projections (lateral, frontal, and axial) of the four different cleft groups superimposed on the control group with UICL. The lateral mean plots are aligned on the *n-s* line and registered at *s*. The frontal mean plots are aligned on the *latero-orbital* line and registered at the center point of that line. The axial mean plots are aligned on a line between the two *tuber points* and registered at the center point of that line. Superimposition of the mean plots for the 2-month-old **a** ICP and UICL groups, **b** RS and UICL groups, **c** UCCLP and UICL groups, and **d** BCCLP and UICL groups

9.2.4 Cleft Lip and Palate (CLP)

Combined clefts of the lip, alveolus, and palate involve structures of both the embryonic primary palate and secondary palate. In Fig. 9.2c the mean craniofacial morphology in 2-month-old unoperated infants with unilateral complete cleft lip and palate (UCCLP) was compared to the control group [33]. The major deviations in the UCCLP group were: decreased posterior length and height of the maxilla; retrognathia of the basal part of the maxilla with relative protrusion of the premaxilla; the width of the maxilla and nasal cavity was markedly increased and the premaxilla deviated to the noncleft side; the mandible was short and retrognathic. Thus, the UCCLP group revealed *bimaxillary retrognathia* combined with a relative protrusion of the premaxilla, which deviated to the noncleft side. In addition, in the UCCLP group the upper airway dimensions were reduced.

Increased width of the midface and nasal cavity was previously reported in *unoperated* UCCLP infants [31] and in *unoperated* adults with UCCLP [49]. Relative protrusion and asymmetry of the premaxilla have also been reported in *unoperated* UCCLP children, adolescents, and adults [5–7, 9, 50, 51]. The relative protrusion and deviation is probably due to overgrowth in the premaxillary-vomerine complex [53, 22, 24], due to the lack of structural integrity of the maxilla on one side. This relative protrusion of the premaxilla explains why we found the measurements *s-n-ans* (*S-N-ANS*) and *s-n-ss* (*S-N-A*) in the infant UCCLP group to be comparable to the values in the control group, despite the fact that the UCCLP group showed significant maxillary retrognathia measured to the basal part of the maxilla.

Dahl et al. [13] and Hermann et al. [39, 40] analyzed facial morphology in 2-month-old infants with unoperated bilateral complete cleft lip and palate from our sample. Fig. 9.2d illustrates the mean facial diagram of the BCCLP group superimposed on the mean facial diagram of the control group. The most obvious features in the BCCLP group were: protrusion of the premaxilla both in relation to the anterior cranial base and in relation to the basal part of the maxilla; the

length of the basal part of the maxilla and posterior maxillary height were decreased; retrognathia of the basal part of the maxilla; markedly increased width of the maxilla and nasal cavity; a short and retrognathic mandible. Thus, the BCCLP group revealed *bimaxillary retrognathia* with a truly protruding premaxilla. In other words, the protruding premaxilla was situated in a totally retrognathic face with a fairly normal sagittal jaw relationship. In addition, the upper airway dimensions were reduced.

The extreme protrusion of the premaxilla is probably the result of marked overgrowth in the premaxillary-vomerine complex secondary to total lack of structural integrity in the region.

For comparison, Mars and Houston [48] and da Silva Filho et al. [58] described groups of adult unoperated patients with BCCLP and found extreme protrusion of the premaxilla and a very convex profile measured as the ANB-angle. No measurements were performed to describe the position of the body of the maxilla. Da Silva-Filho et al. [56, 58] also found the mandible to be short and retrognathic and discussed whether this finding was related to the primary anomaly or if it was caused by secondary functional adaptations.

The retrognathia of the basal part of the maxilla, and the short and retrognathic mandible found in our sample are, in our opinion, variations intrinsically associated with the cleft of the secondary palate as discussed above.

9.3 Discussion and Conclusions

The Danish study of craniofacial morphology in untreated cleft infants is the hitherto most comprehensive and well-controlled, since it covers a whole population, which is homogeneous and in which central registration of clefts has been carried out for more than 65 years; a registration which has been shown to be highly reliable and nearly complete. Furthermore, all cleft infants are surgically treated at one hospital by one surgeon using the same techniques. All infants were examined with state-of-the-art three-projection cephalometry using the hitherto most comprehensive cephalometric analysis covering all craniofacial regions and the methods were validated. The study included more than 600 children, and even after breakdown into subgroups, the sample sizes were adequate for statistical testing (except maybe for the RS group). Based on these facts, the findings related to the infant craniofacial morphology at 2 months of age, prior to any surgical or orthopedic treatment, must be considered to represent the “true” malformation, primarily caused by intrinsic factors.

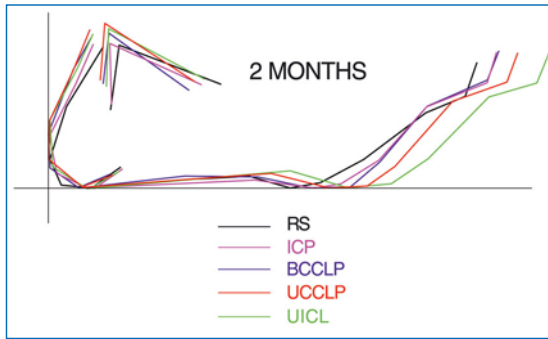


Fig. 9.3. Mean plots of the mandible in the RS, ICP, BCCLP, UCCLP, and UICL groups. Superimposition was made on the mandibular line (ML) registered at pogonion (pg)

In Table 9.1 the most important findings in the primary anomaly in the Danish infants with RS, ICP, BCCLP, and UCCLP are given, revealing a rather clear pattern. The findings support the suggestion of Dahl [12] and others, that facial clefts should be classified based on the embryonic facial development, i.e., into clefts involving the primary palate only (CL), clefts involving the secondary palate only (CP), and clefts involving structures of both the primary and the secondary palate (CLP). The postnatal facial morphology in these groups differs greatly. Infants with cleft of the secondary palate, with or without cleft of the primary palate, shared a number of characteristic morphological traits when compared to the norm: decreased posterior length of the maxilla; maxillary retrognathia; decreased posterior height of the maxilla; increased width of the maxilla and the nasal cavity; decreased length of the mandible; mandibular retrognathia; and reduced size of the pharyngeal airway. As seen from Table 9.1 and Fig. 9.3, the mandibular involvement was most pronounced in the RS group followed by the ICP and BCCLP groups, and, finally, the UCCLP group. A similar pattern was observed for the reduced size of the pharyngeal airway. As for the maxilla, the increased width of the maxilla and the nasal cavity was most pronounced in the groups with clefts of both the secondary and the primary palate, i.e., BCCLP and UCCLP. None of these groups showed decreased total length of the maxilla or retrognathia of the maxilla when measured to the premaxilla; the reason for this being a true and relative protrusion of the premaxilla, respectively.

In conclusion, a short and retrognathic mandible was a constant finding in infants with cleft of the secondary palate. The reduction in size of the pharyngeal airway in infants with cleft of the secondary palate was clearly related to the short and retrognathic mandible, being most severe in the RS group, which

had the added effect of a reduction in the cranial base angle. But, in principle, all four groups had restricted upper airways as part of the primary anomaly. The increased width of the maxilla and nasal cavity was most pronounced in the groups which also had cleft of the primary palate (UCCLP and BCCLP). The UCCLP group was also characterized by relative protrusion of the premaxilla which was positioned asymmetrically, deviating to the noncleft side, whereas in the BCCLP group the premaxilla showed true protrusion both in relation to the basal part of the maxilla (the lateral segments) and to the anterior cranial base. On average, the premaxilla was found to be positioned in the midline in this group, although most of the individual cases showed some degree of asymmetry. The protrusion of the premaxilla is suggested to be secondary to the primary anomaly of clefting, allowing for overgrowth in the premaxillary-vomerine complex, due to partial or total lack of anatomical integrity in the region.

It has been the aim of this chapter to summarize our findings about the intrinsic variations in facial morphology associated with the different types of cleft malformations to form a basis for valid estimations of the amount of surgical iatrogenesis, especially to the maxillary development, introduced by different surgical procedures and regimes, including the timing of treatment. In Fig. 9.4, the growth changes of the craniofacial skeleton from 2 to 22 months of age in the UCCLP group has been compared to the UICL group (control group) using color-coded surfaces on a 3D CT-model. In both groups the cleft lip was surgically closed just after the examination at 2 months of age using a Tennison procedure. In the UCCLP group, the anterior part of the palate was closed with a vomer flap at the same time. The method of producing the illustrations will be given below.

9.3.1 Intuitive Visualization of the Location of Growth Differences

Cephalometric measurements in three projections provided growth vectors at each of the 279 (230 skeletal and 49 soft tissue) anatomical landmarks. The growth vectors, computed as the vector difference between corresponding landmark locations at the ages 2 and 22 months, respectively, after alignment to a common coordinate system [35], have been used to form average growth patterns previously shown in Hermann et al. [34, 33] (UICL, UCCLP) and Hermann et al. [41] (UICL, BCCLP). Results of comparisons of growth between the UCCLP and the BCCLP groups, respectively, and the control group (UICL) have been shown as color-coded average growth patterns in Hermann et al. [33, 34] (UCCLP vs. UICL)

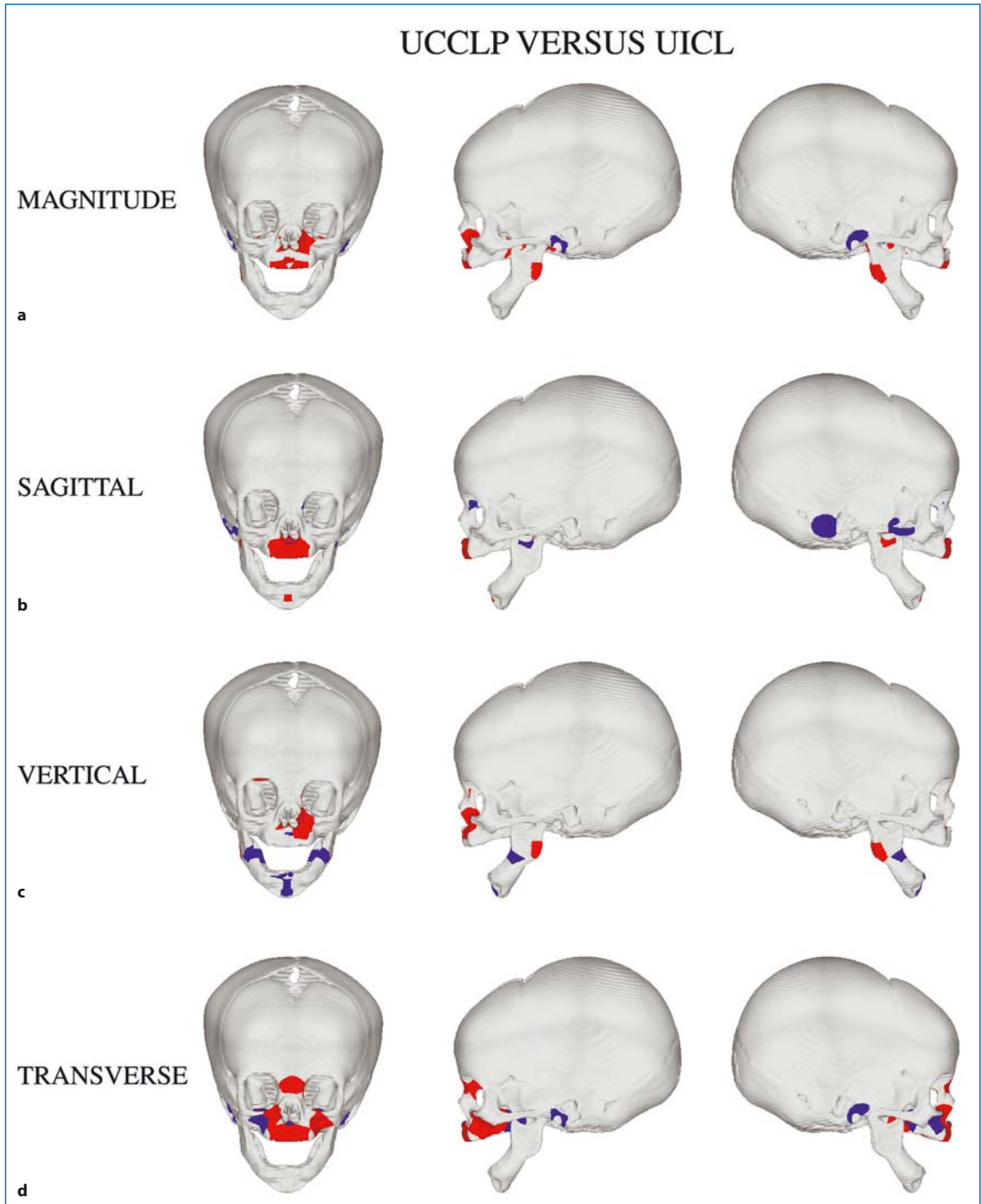


Fig. 9.4 a–d. 3D visualization of locations of growth differences. Locations where UCCLP growth differs significantly ($p < 0.01$) from UICL growth (2–22 months of age) are colored red (UCCLP < UICL) or blue (UCCLP > UICL). The surface reconstruction shown is of a noncleft subject of comparable

age, and is used solely for illustration. Cleft side is on patient's left in the figures. Locations of differences in the **a** magnitude of growth, **b** sagittal, **c** vertical and **d** transverse growth components are shown

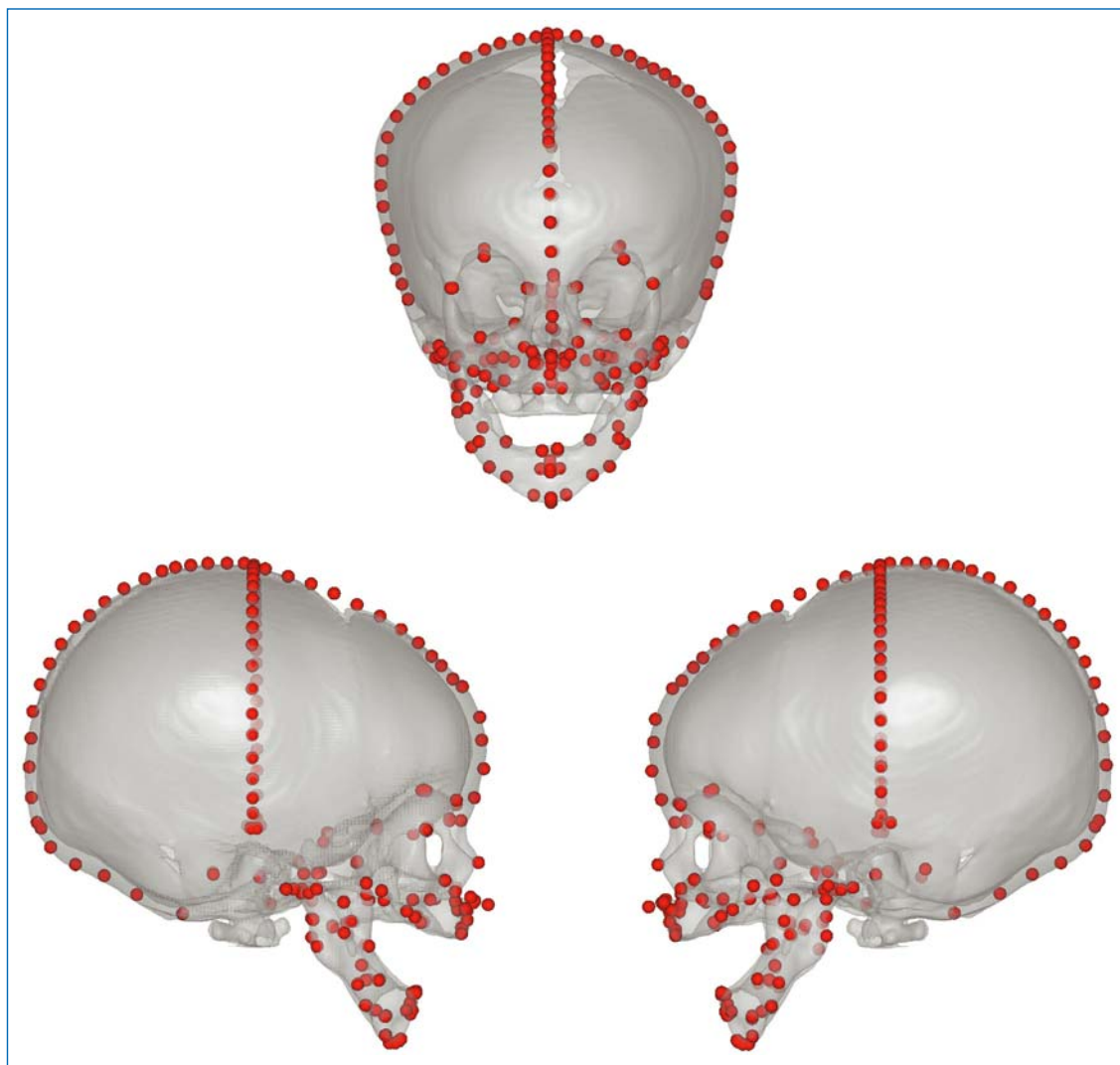


Fig. 9.5. 3D landmark locations corresponding to the skeletal landmarks used in the three-projection cephalometric analysis as well as for creating the color-coded surfaces in Fig. 9.4

and Hermann et al. [41] (BCCLP vs. UICL). These color-coded growth diagrams disclosed the locations of significantly different growth (1%, 5%, and 10% levels) in the study group when compared to a reference group, and the diagrams were shown separately for each of the 3 projections (lateral, frontal and axial), as well as for the growth magnitude and the two growth directions (x and y in each of the projections, respectively). In order to facilitate the effective comprehension of these diagrams, the locations of significant difference are color-coded onto the surface of a skull reconstructed from a CT scan of a single (non-cleft) infant. As an example, Fig. 9.4 shows such color-coded surfaces for the comparison of the UCCLP with

the UICL (control group). The color-coded surfaces were created by landmarking the 3D CT scan of the single noncleft infant at locations corresponding to the 230 skeletal cephalometric landmarks and color coding the surface in the vicinity of each landmark by a color corresponding to the significance of the growth difference. The landmark locations are shown in Fig. 9.5. A color table was chosen such that colors signify Student's t-test p values smaller than 0.01. Blue colors correspond to locations where the study group exhibits larger growth than the control group, while the opposite is the case at locations colored red. Regions without any significant differences between the two groups remained gray. In the UICL and UCCLP

groups the frontal and axial projection data were mirrored in order to have all clefts on the left side. Accordingly, the cleft is on the patient's left side in Fig. 9.4. The spatial extent of colored surface area in the vicinity of a landmark was governed by the distance to its closest landmark, and a maximum extent (spherically from landmark position) was chosen as 40 mm. Color-coded skulls are shown for differences in growth magnitude, as well as for each of the three growth directions (sagittal, vertical and transverse). The colors for sagittal growth differences were computed from the x-component of the growth vectors in the lateral cephalometric projection and the y-component of the growth vectors in the axial projection. The colors for vertical growth differences were computed from the y-component of the growth vectors in the lateral projection and the y-component of the growth vectors in the frontal projection. The colors for transverse growth differences were computed from the x-component of the growth vectors in the frontal projection and the x-component of the growth vectors in the axial projection. The method of color coding has previously been described and applied for visualization of the growth differences between UCCLP and UICL in Darvann et al. [16].

Secondary to surgical closure of the lip at 2 months of age in the UCCLP group we found that the premaxilla was molded into place, demasking the intrinsic maxillary retrognathia and leading to a normal sagittal jaw relationship at 22 months of age. Maxillary growth was, besides the premaxillary molding, characterized by smaller vertical growth on the cleft side and reduced transverse development, which could probably be related to the effects of surgery. The amount of mandibular growth was similar in the two groups. However, the direction of growth was slightly more vertical in the UCCLP group. This growth pattern was probably related to the intrinsic pattern of mandibular development. Otherwise, craniofacial growth seemed to be very similar in the two groups.

We found that surgery to the lip and anterior part of the hard palate at 2 months of age in UCCLP subjects seemed to influence the development of the maxillary complex, as observed at 22 months of age, in a number of beneficial ways: the premaxilla was no longer relatively protruding, and it was less asymmetric; the nasal septum deviated less toward the noncleft side; the width of the nasal cavity and the posterior part of the maxilla became relatively more normal; and the transverse position of the lateral maxillary segment on the noncleft side was closer to normal. The posterior height of the maxilla was, however, still reduced to the same degree; the mandible was still short and retrognathic to the same degree; and bimaxillary retrognathia was still present. The only

iatrogenic effect observed was that the lateral maxillary segment on the cleft side had become displaced toward the midsagittal plane anteriorly, resulting in a much too narrow dental arch at the level of the deciduous canine [35].

It is noteworthy that several studies of older, *unoperated* UCCLP children and adults find the maxillary prognathism to be within normal limits or even increased when compared to normative data [9, 48, 50, 51]. All these studies, however, only measure maxillary prognathism to the A-point or to the point ANS, both located in the relatively protruding premaxilla. Ortiz-Monasterio et al. [50] concluded based on their findings in unoperated adults with UCCLP, that: "The embryonic factor responsible for the facial cleft does not interfere with maxillary growth. This evidence leads us to believe that growth defects of the middle third of the face so frequently seen are caused by early or repeated and aggressive surgery." We disagree somewhat with this conclusion. Based on our studies of infants with UCCLP, it would seem that maxillary retrognathia in this group is part of the intrinsic variations associated with the cleft malformation of the secondary palate. In the unoperated infant and the unoperated adult the maxillary retrognathia is, however, partly masked by relative protrusion of the premaxilla, secondary to overgrowth in the premaxillary-vomerine suture. Surgical closure of the lip at 2 months of age molds the premaxilla back into place, demasking the maxillary retrognathia. Thus in the 22-month-old lip-operated UCCLP group, it is our opinion that the bimaxillary retrognathia illustrates the facial type characteristic of the group rather than an iatrogenic effect of cleft surgery [34, 35]. Thus, we do not consider the maxillary retrognathia observed at 22 months of age as the result of surgical iatrogenesis, rather we believe it represents a normalization of the "intrinsic facial type" characteristic of subjects with UCCLP; and at 22 months of age the face is still harmonious with a normal sagittal jaw relationship. We have, at this point in time, not re-examined the sample at older ages, and can, therefore, not comment on facial growth and signs.

In conclusion, we are not arguing that cleft surgery does not lead to disturbed maxillary development during the growth period. But we are suggesting that subjects with cleft of the secondary palate have a special "intrinsic" facial type, primarily characterized by bimaxillary retrognathia and increased maxillary width. We are speculating that this facial type could be a "liability factor" increasing the probability of CP or CLP [33, 34]. Finally, we suggest that when outcome of cleft surgery in CLP subjects is evaluated at adolescence or adulthood, comparisons should not be made to normal standards, but rather to the adolescent and adult morphology seen in CP subjects.

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10 Facial Growth and Morphology in the Unoperated Cleft Lip and Palate Subject: The Sri Lanka Study

MICHAEL MARS

Studies of subjects with unoperated clefts of the lip and palate have been undertaken for the past 80 years [1–29].

Adults with unoperated clefts of the lip and palate provide the ideal control group for investigators studying the natural history of facial growth and morphology in these subjects. The absence of surgical intervention provides an opportunity to study the outcome of facial growth, morphology, and speech, using an absolute comparative baseline. An evaluation of the intrinsic versus the potential iatrogenic influences can be separately analyzed, when this group is compared to conventionally operated subjects. They also highlight the discrepancies in health care provision between the wealthy western world and the developing/least developed world.

Withdrawal of surgery for research purposes would be unethical where such facilities are available. Likewise, assignment to surgical or nonsurgical management programs on a prospective random allocation basis could not be permitted. For these reasons, studies on the unoperated subject have been made on individuals from the developing world, where surgery may not be readily available. This has led to many of the limitations listed below being evident in such studies. Additional limitations have been imposed by the authors themselves, using inappropriate management of the material collected.

The difficulties experienced by research workers in the field of cleft lip and palate are seen in the limitations of many studies. These may be summarized as follows:

1. Small sample size
2. Wide age distribution
3. A narrow age distribution of very young subjects
4. Mixtures of unoperated, partially operated, late operated and early operated subjects
5. Mixtures of subjects from different cleft types
6. Males and females grouped together
7. No controls from the normal population

8. Controls of treated patients from the same population seldom available (applicable to studies of unoperated versus operated subjects)
9. Few postoperative follow-up studies where late surgery has been performed

Unquestionably, the major problem is that of small sample size, which has been responsible for the inappropriate handling of the available data.

The general consensus in the literature relating to the unoperated UCLP subject suggests that they have the potential for near normal facial growth. However, serious limitations are obvious in the analysis of the data, especially by attempts to increase the sample size by pooling nonhomogeneous groups. Despite limitations of all previous studies into facial growth in the totally unoperated cleft lip and palate subject, the results suggest that facial growth proceeds reasonably well. Midface depth, the parameter of most concern in CLP, is not compromised in unoperated subjects.

10.1 Sri Lankan Cleft Lip and Palate Project

The Sri Lankan Cleft Lip and Palate Project has developed as the largest multidisciplinary surgical and research program concerned with the unoperated cleft lip and palate subject [30]. Since 1984, extensive records have been collected on over 1,000 subjects, on whom 820 operations have been performed. There have been 13 visits from 1984 to 2002. The aims have always been threefold: treatment, teaching, and research based on a multidisciplinary team. Over 50 professionals have been involved, many having attended on over 10 visits.

The unresolved conflicts of opinion regarding the aetiology of facial growth distortion in repaired cleft lip and palate subjects were the main reasons for the establishment of this Project. The possibility of

Table 10.1. Unoperated groups

	UCLP				BCLP				ICP			
	Young		Mature		Young		Mature		Young		Mature	
	M	F	M	F	M	F	M	F	M	F	M	F
Sample size	15	8	21	11	5	6	5	8	8	15	9	9
Age range	13–20	14–18	21–47	19–43	15–19	14–19	21–49	21–55	13–20	13–18	21–44	19–43
Mean age	15.5	15.8	28	26.8	17.2	16.6	30.2	32	N/k	N/k	N/k	N/k

Table 10.2. Control group

	Young		Mature	
	Male	Female	Male	Female
Sample size	21	58	24	16
Age range	14–19	14–18	20–30	20–30
Mean age	15.9	16.2	25.6	27.5

addressing these questions by studying “nature’s experiment” on hundreds of unoperated and late-operated subjects of all ages from birth to old age was unique. Such an opportunity is rare. Investigators usually have to resort to animal experiments in order to provide sufficient numbers in a controlled manner [31, 32].

This chapter, which deals with totally unoperated subjects from Sri Lanka presents a descriptive account of facial morphology, a cephalometric analysis (UCLP, BCLP, and ICP groups), a GOSLON yardstick analysis (UCLP group), and a reflex microscopic analysis of dental study models (UCLP and ICP groups). The material comprises 55 UCLP, 23 BCLP, and 41 ICP subjects (Table 10.1), and 119 healthy control Sri Lankan subjects (Table 10.2). All unoperated subjects described in this chapter were over 13 years of age when they first presented.

Because the onset of puberty is some two to three years later in Sri Lanka than in the West, it was necessary to separate pre- and postpubertal groups [33]. Further, there are significant differences in the timing of the onset and termination of puberty between males and females. This, and the fact that there are some significant differences in facial morphology between the sexes, has necessitated their separate analysis.

10.1.1. Controls

One hundred nineteen healthy adult Sri Lankan non-cleft subjects provided Control data. These comprised 40 male medical students and 79 female hospital nurses and medical students. The Controls were aged between 20 and 30 years of age (Figs. 10.1, 10.2).

10.1.2 Records Collected for Study

Lateral skull radiographs and dental study models of the unoperated subjects comprise the material for cephalometric and Goslon Yardstick analyses.

10.1.3 Radiographs

The lateral skull radiographs were taken in a cephalostat sited in a private sector hospital in Galle. The same design of cephalostat was used in Kandy and Galle on all subsequent visits.

In 1984, the protocol for setting up the radiographic apparatus was established and supervised for each exposure by the author. In subsequent visits, all orthodontic members of the Team, after training, took turns in placing the subjects in the cephalostat and supervising all exposures.

Great care was taken at each radiographic session to ensure that the anode to midsagittal distance was precisely 152.5 cm (the Imperial measurement of 5 ft) and the midsagittal plane to film distance was 16 cm. The central ray was arranged at right angles to the sagittal plane. This was determined by an electric light source within the anode housing, casting a superimposed shadow of both ear rods on a sheet of white paper which was attached to the x-ray film cassette. These measures ensured reproducible skull radiographs on all occasions with consistent magnification error. The patients were posed with the teeth lightly occluded in maximal intercuspation and the Frankfort plane parallel to the ground.

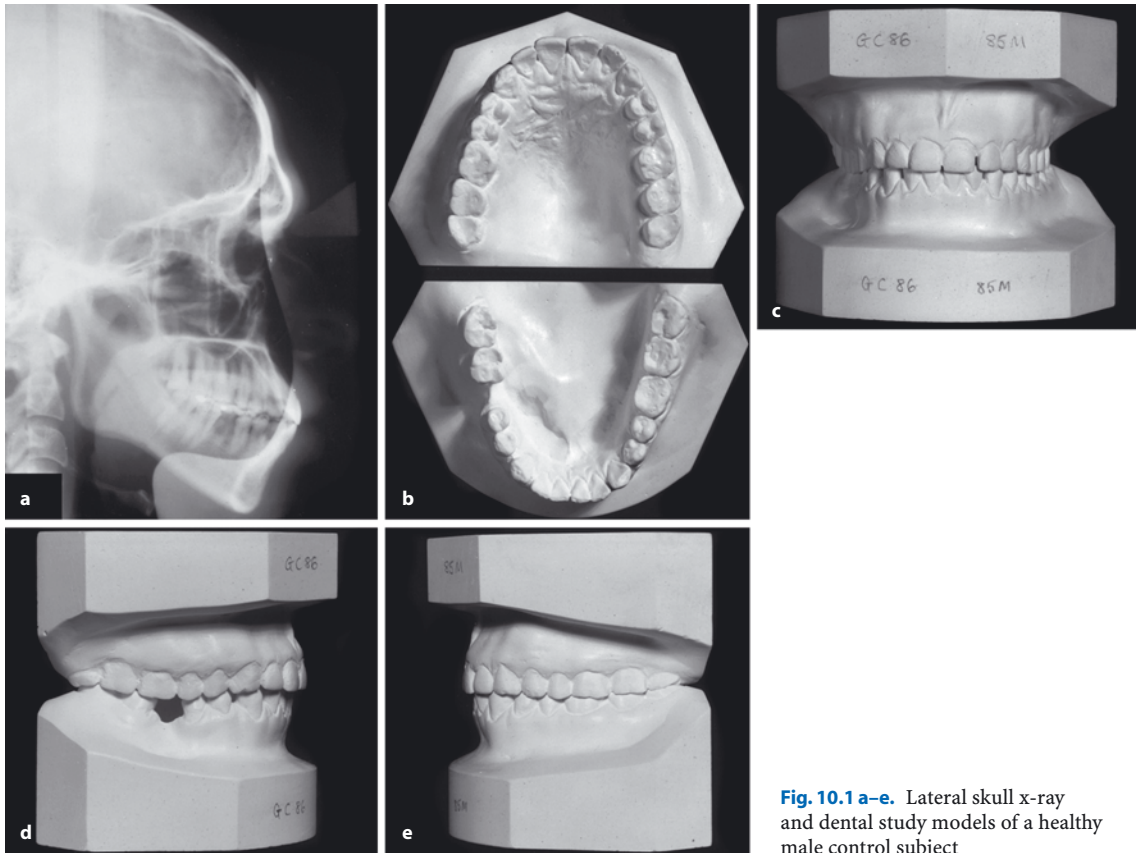


Fig. 10.1 a–e. Lateral skull x-ray and dental study models of a healthy male control subject

Table 10.3. Mature unoperated subjects cephalometric results (UCLP, BCLP, and ICP)

	Controls Mature		UCLP Mature		BCLP Mature		ICP Mature	
	M	F	M	F	M	F	M	F
Ba_N Cranial Base	112.5±2	108±2	110±2	110±4	102.3±7	100.2±8	108.4±5	100.3±5
SNA Maxillary protrusion	85.4±3	85±3	85±2	86±4	90.3±10	90.6±8	83.2±6	83.4±4
SNANS Basal maxillary protrusion	87.1±3	87.1±3	84±2	86±4	94.6±10	94.5±8	84.7±6	85.3±5
Ans-Ptm Palatal Length	57.8±5	53.1±4	54±2	51±2	61.6±6	61.3±6	50.3±6	49.2±5
SNB Mandible protrusion	82±4	83.4±3	80±2	81±3	81.6±5	81.7±6	81.3±5	81.3±4
AR-TGO Ramus height	57.5	50.0	51.3	48.9	48.5±4	47.8±41	48.6	47.1
S_N_ANS Upper face height	54.5	51.5	52.6	47.5	46.6±3	45.5±2	49.2	45.6
N-S-Ptm Upper post face height	44.0	40.3	38.3	37.0	33.5±4	31.2±3	39.7	33.4

Temperatures were always around 30°C, and the humidity was often near 100%, but this did not adversely affect the quality of the films, which were immediately processed in wet tanks. They were dried

and suspended from wire “washing lines” strung across the main x-ray room. Patients were detained until after their x-ray had been examined, in case a repeat x-ray was required (Fig. 10.3, Table 3).

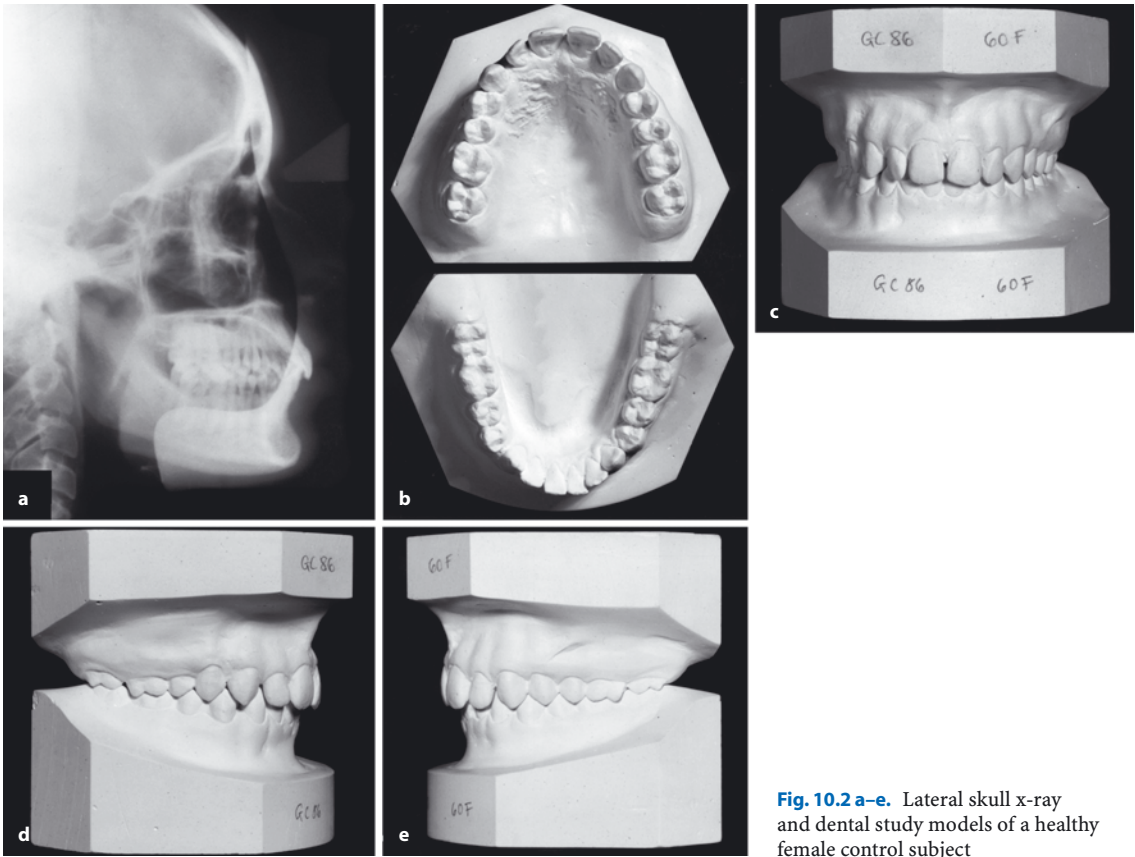


Fig. 10.2 a–e. Lateral skull x-ray and dental study models of a healthy female control subject

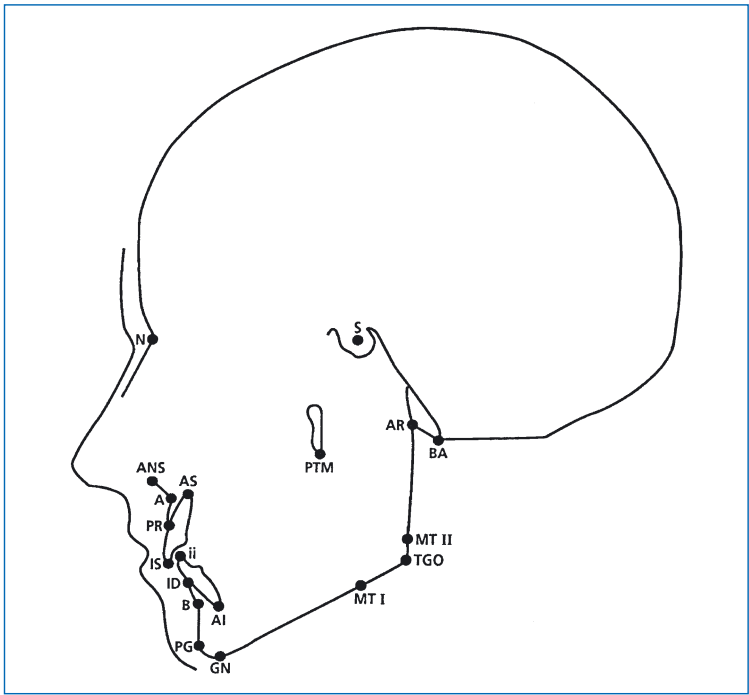


Fig. 10.3. Digitized points used for cephalometric analysis

10.2 Unoperated Unilateral Cleft Lip and Palate

The most striking feature in the unoperated unilateral cleft lip and palate subject is the protrusion of the upper labial segment. Subjects present with large overjets, proclined upper incisors, eversion of the major segment and a mild contraction of the lesser segment in the anterior region. Buccal cross-bites are rare. Cephalometric analysis displays normal cranial base measurements and protrusion of the maxilla relative to the mandible. No cases presented with maxillary retrusion (a common feature in the operated case). Figures 10.4 and 10.5 illustrate typical examples of the facial appearance and dental study models of unoperated unilateral cleft lip and palate cases.

10.2.1 Dental Study Models

Impressions were taken in alginate material with the patients sitting upright on a wooden chair. Bite registration was made using conventional denture wax. In 1984, the author and accompanying oral surgeon cast the impressions in white dental plaster in the hotel bathroom after the day's work. This process was fraught with difficulty because dental plaster sets extremely rapidly in the tropics.

On all subsequent visits, a local Sri Lankan technician was employed. Impressions were cast in dental stone on the day that they were taken, incorporating the tooth bearing and important anatomical areas. Care was taken to include all anatomically visible structures. This necessitated modifying aluminium

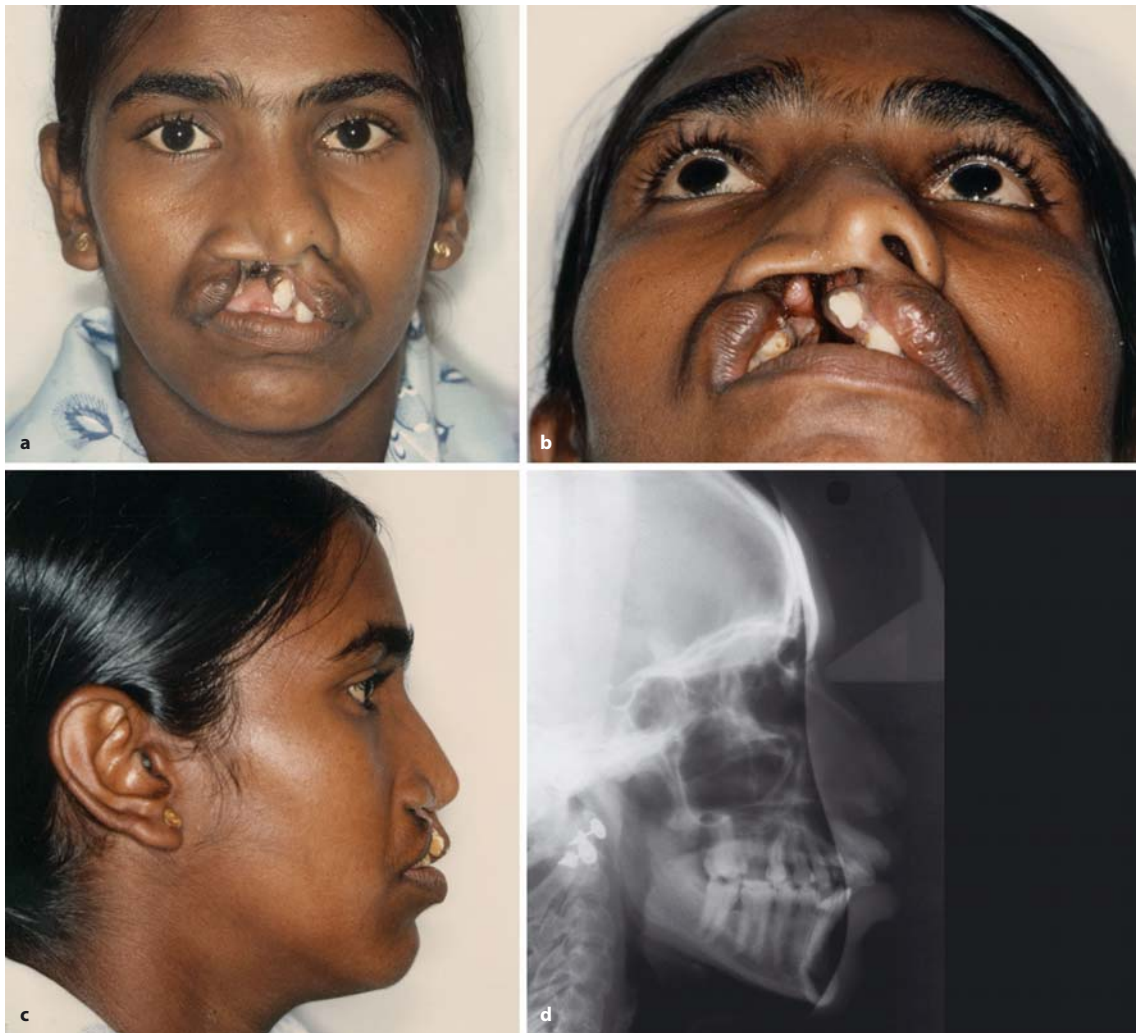


Fig. 10.4 a–j. Adult female subject with complete unilateral cleft of the lip and palate

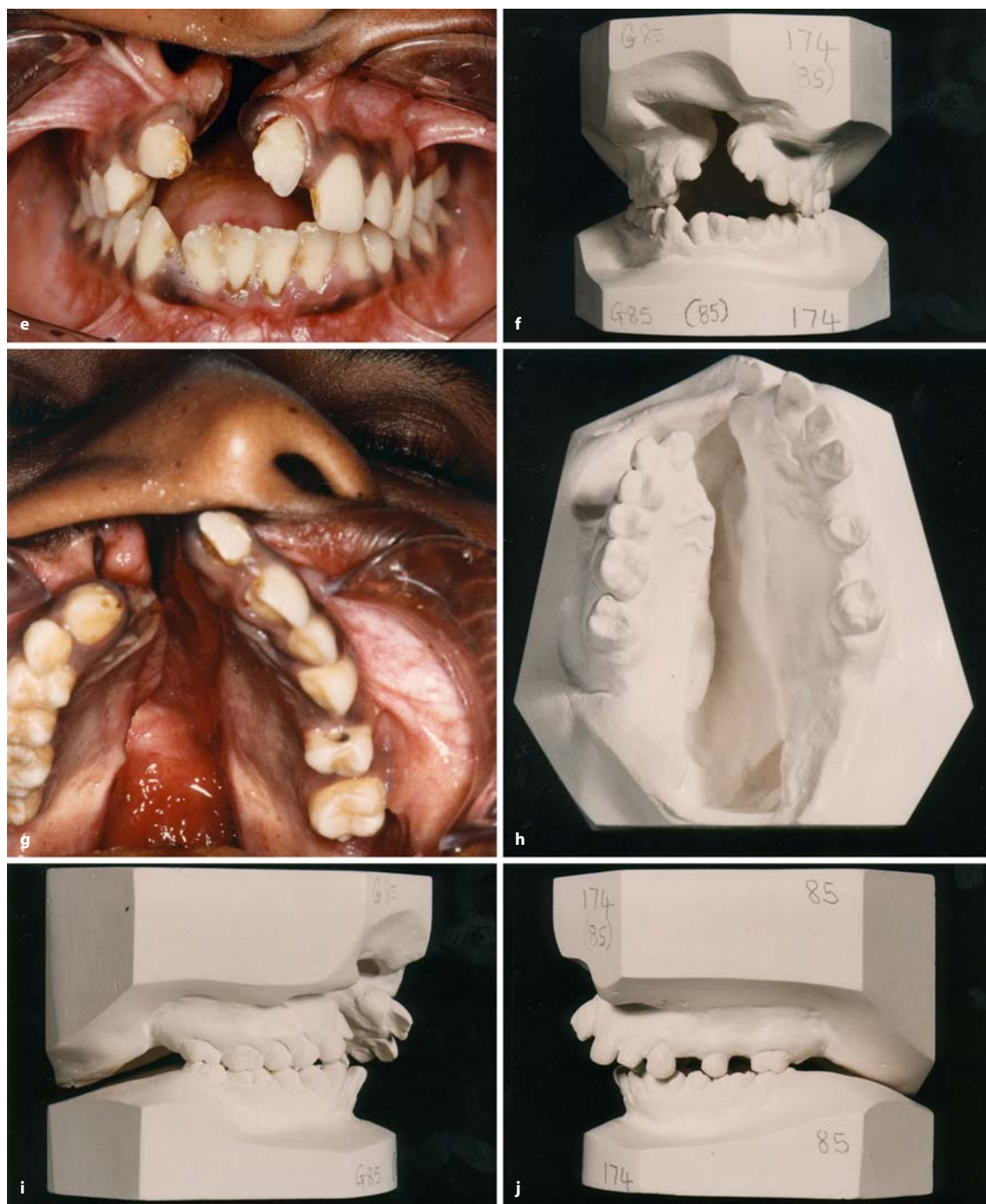


Fig. 10.4 a–j. (continued)

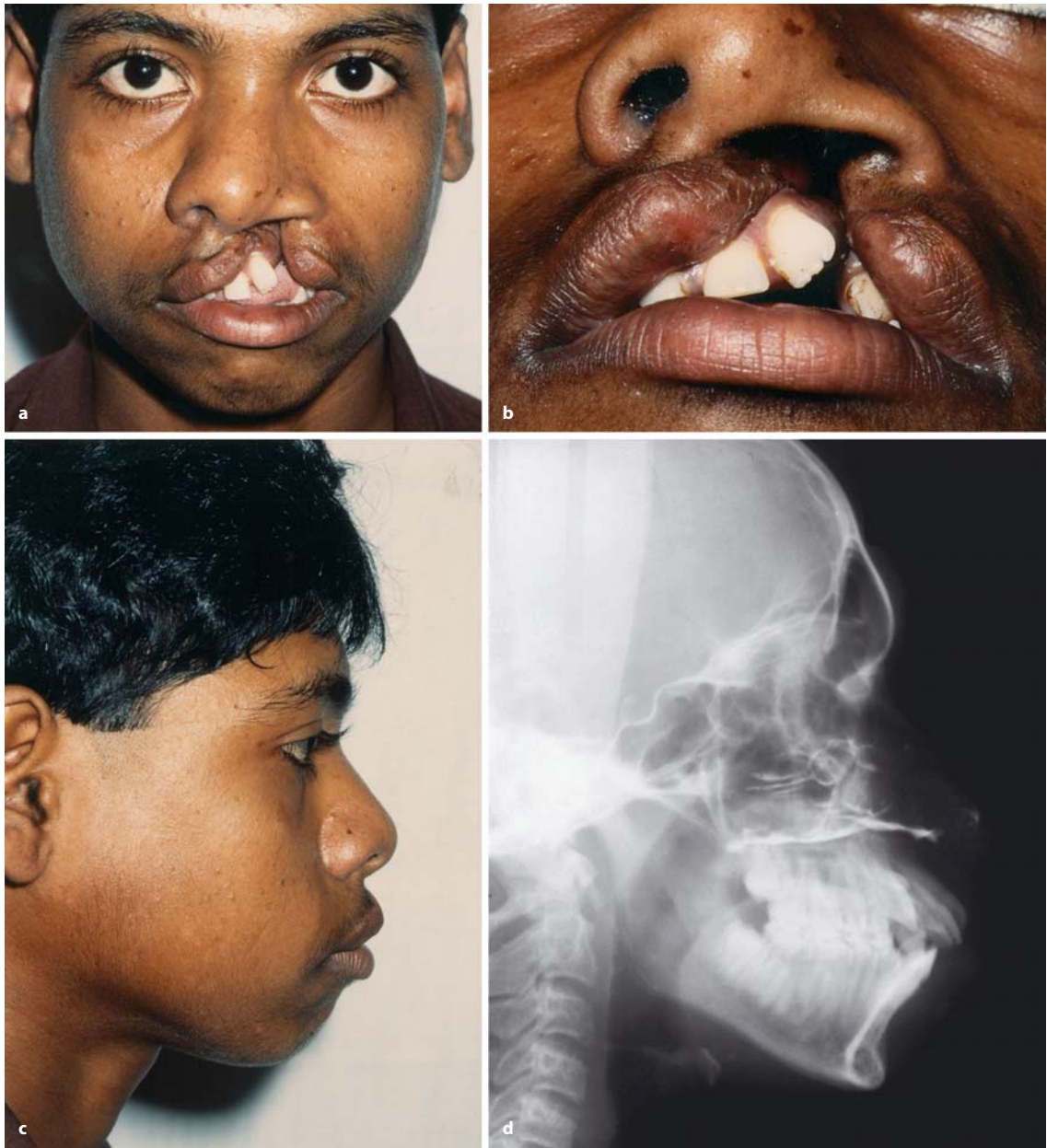


Fig. 10.5 a–j. Adult male subject with complete unilateral cleft of the lip

trays with white wax rims. Huge volumes of alginate were used on some of the adults with wide-open clefts, and included impressions of the middle and inferior turbinate bones, the entrance to eustachian tubes, and the posterior wall of the pharynx. Some impressions utilized 13 scoops of alginate powder. None of the patients vomited. Figure 10.6 shows algi-

nate impressions with extension beyond the inferior turbinates, the entrance to the eustachian tubes, and the posterior wall of the pharynx.

Final orthodontic trimming and basing of the models was performed at the laboratory in the Maxillofacial and Dental Department in the Hospital for Sick Children, Great Ormond Street, London.

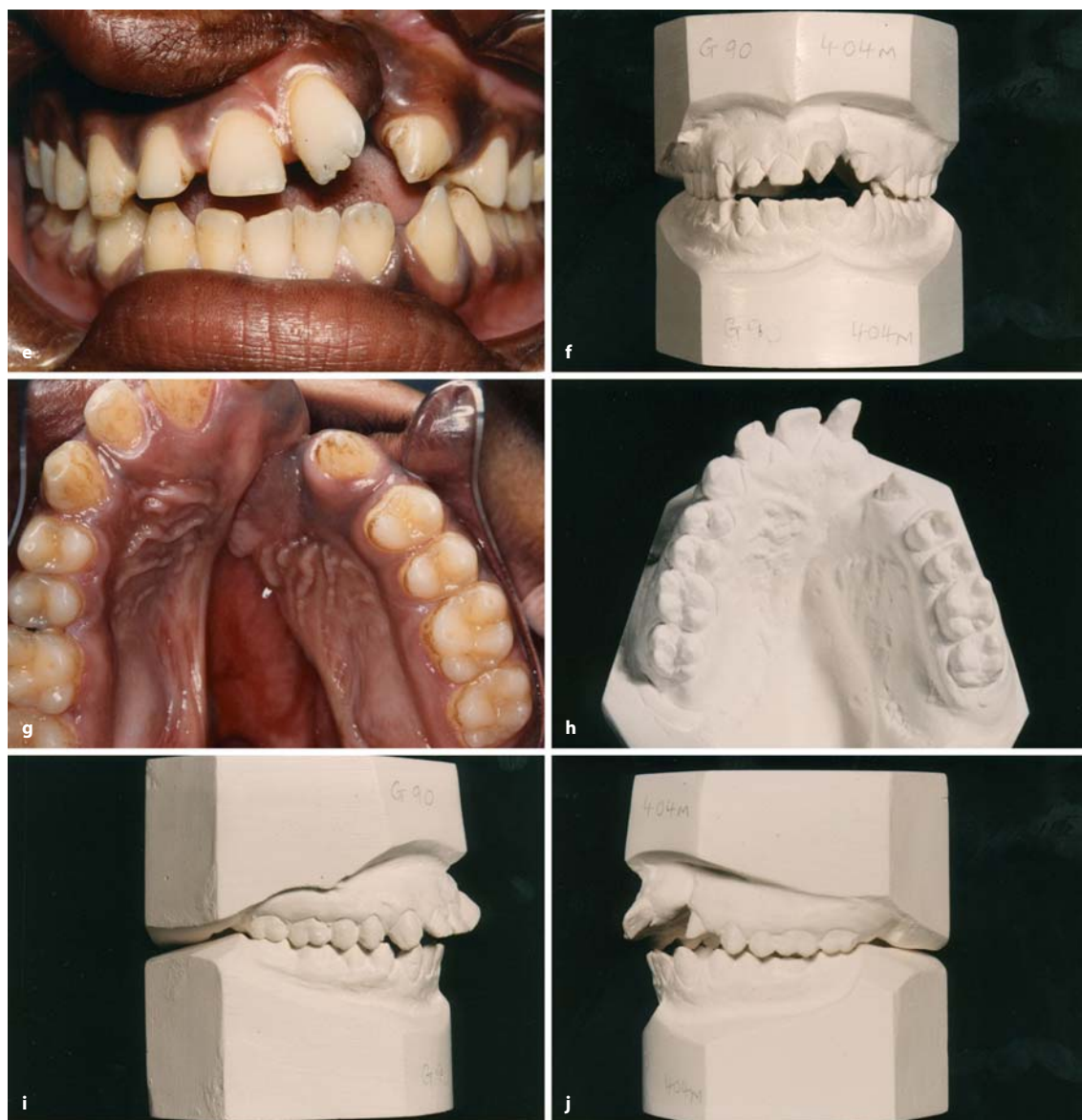


Fig. 10.5 a–j. (continued)

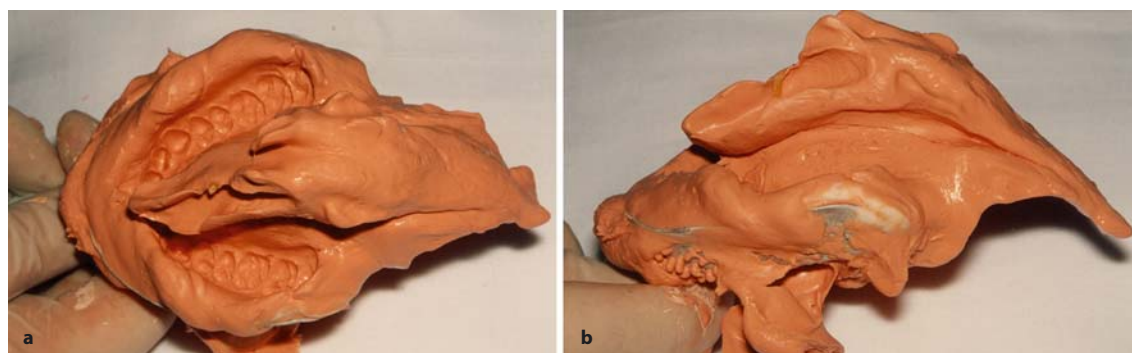


Fig. 10.6 a, b. Upper arch alginate impressions, maximally extended beyond the inferior turbinates, the entrance to the eustachian tube and the posterior wall of the pharynx

Fig. 10.7. GOSLON Grouping, Sri Lankan Unoperated UCLP. n = 51; 98% in Excellent (1) or Good (2) arch relationships

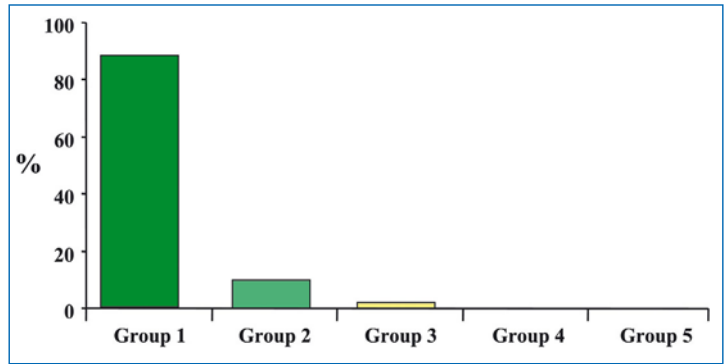
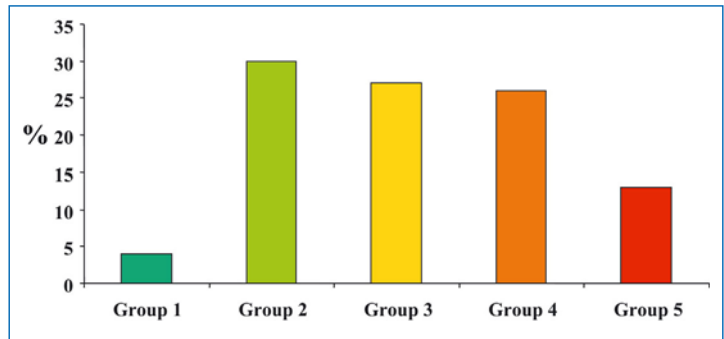


Fig. 10.8. UK CSAG Study 1998. GOSLON CSAG (Clinical Standards Advisory Group) GOSLON results of the whole of the UK



10.2.2 The GOSLON Yardstick

The GOSLON Yardstick [34] a London/Oslo consensus of 5 groups of increasing deformity in dental arch relationships was applied to 49 unoperated cases for whom cephalometric analysis had been performed. Two additional cases where study models, but not lateral skull radiographs, were available have been included.

“UNOPS” in GOSLON STUDY(UCLP) [51]

<i>Younger:</i>	Male	Female
	13	8
<i>Mature:</i>	Male	Female
	19	11

The ranking was performed by the author. Previous reports have demonstrated the reproducibility of the author's assessments, and the robustness of the Yardstick in the discrimination between differences in samples from different centres [35, 36]

The GOSLON yardstick results showed 98% of the cases in group 1 or 2 (excellent or very good arch relationships) and no cases in groups 4 or 5 (Fig. 10.7).

Because the yardstick is not a direct measure of skeletal morphology or growth, it is possible to pool males and females over 13 years in a single group.

These results are in marked contrast to those of any centre worldwide examining operated patients, where only a very small minority of patients are found in group 1 (Fig. 10.8).

10.2.3 Unilateral Cleft Lip and Palate Study Models Analysis by Reflex Microscope (Fig. 10.9)

10.2.3.1 Arch Widths

The cleft group was narrower in the second molar region by a mean of 1.6 mm and narrowed progressively to a maximum mean deficit of 5.0 mm in the canine region, resulting in the arches being more V shaped. There were no differences according to sex.

10.2.3.2 Tooth Widths

There were significant differences in tooth size between the cleft group and the controls: the teeth of the cleft group were consistently smaller and the largest differences were in the central incisors. No significant differences were found in comparisons of the cleft and noncleft sides, sexes, or side to side within each group.

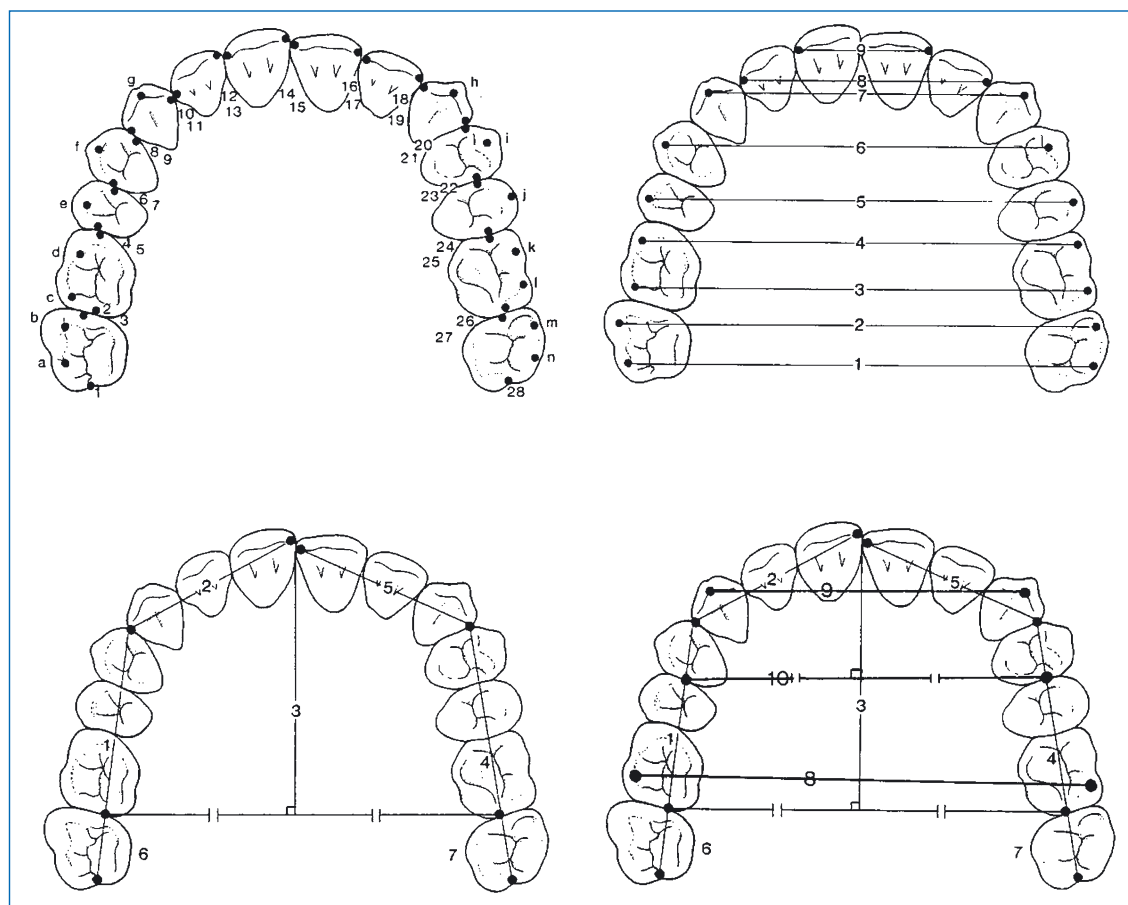


Fig. 10.9. The digitized points, chord lengths, and arch widths used in the reflex microscopic analysis of study model

10.2.3.3 Chord Lengths

No significant differences were found in chord lengths.

10.2.3.4 Crossbites

There was a higher prevalence of crossbites in the cleft subjects (8 cases) than in the control group where there were none.

10.2.3.5 Overjet

The overjet was much larger in the cleft group. The mean overjets in the cleft group was 8.2 mm compared to 3.7 mm in the controls

10.2.3.6 Missing Teeth

The percentage of missing teeth was higher in the cleft group; the most commonly missing teeth in the cleft group were the maxillary lateral incisors.

10.2.3.7 Crowding

There was no crowding in the buccal segments of either the control group or the cleft group. In fact, all dental arches examined were very well aligned and some spacing was generally evident.

10.2.4 Summary of Reflex Microscope Findings on Study Models: UCLP [15]

Tooth size and arch widths that were found to be smaller in the unilateral cleft lip and palate subjects than in the control group would suggest that there is some degree of primary hypoplasia in this group. However, these differences are small and would not account for the gross maxillary retrusion frequently reported in surgically repaired unilateral cleft patients. Even with this small primary hypoplasia, overjets in the cleft group are larger than the controls and the prevalence of crossbites is surprisingly low. These results concur with the cephalometric findings of a smaller retrusive mandible in the unilateral cleft and palate subject.

These features demonstrate the intrinsic potential for these subjects to grow relatively normally with minor distortions around the cleft site itself where the dentition is unrestrained because of the disrupted musculature. There are, however, some intrinsic growth deficiencies, particularly a shortened ramus

height, and reduced upper anterior midface height and upper posterior face height.

10.3 Unoperated Bilateral Cleft Lip and Palate

Unoperated bilateral cleft lip and palate cases demonstrate massive protrusion of the premaxilla with gross proclination of the upper incisors. Interestingly, they show significantly smaller cranial base lengths. There are intrinsic growth deficiencies; a shortened ramus height, and reduced upper anterior midface height and upper posterior face height. The initial characteristics of the newborn bilateral cleft lip and palate subject persist during growth. The prominent premaxilla, wide alar bases, often laterally deviated premaxillary segment, short columella, and rudimentary prolabium with no muscle attachment are characteristic features. Figures 10.10–10.12 illustrate the typical examples of the facial appearance and dental study models of unoperated bilateral cleft lip and palate cases [37].



Fig. 10.10 a–d. Mature male BCLP, note lateral segments have moved to the midline excluding the premaxilla anteriorly

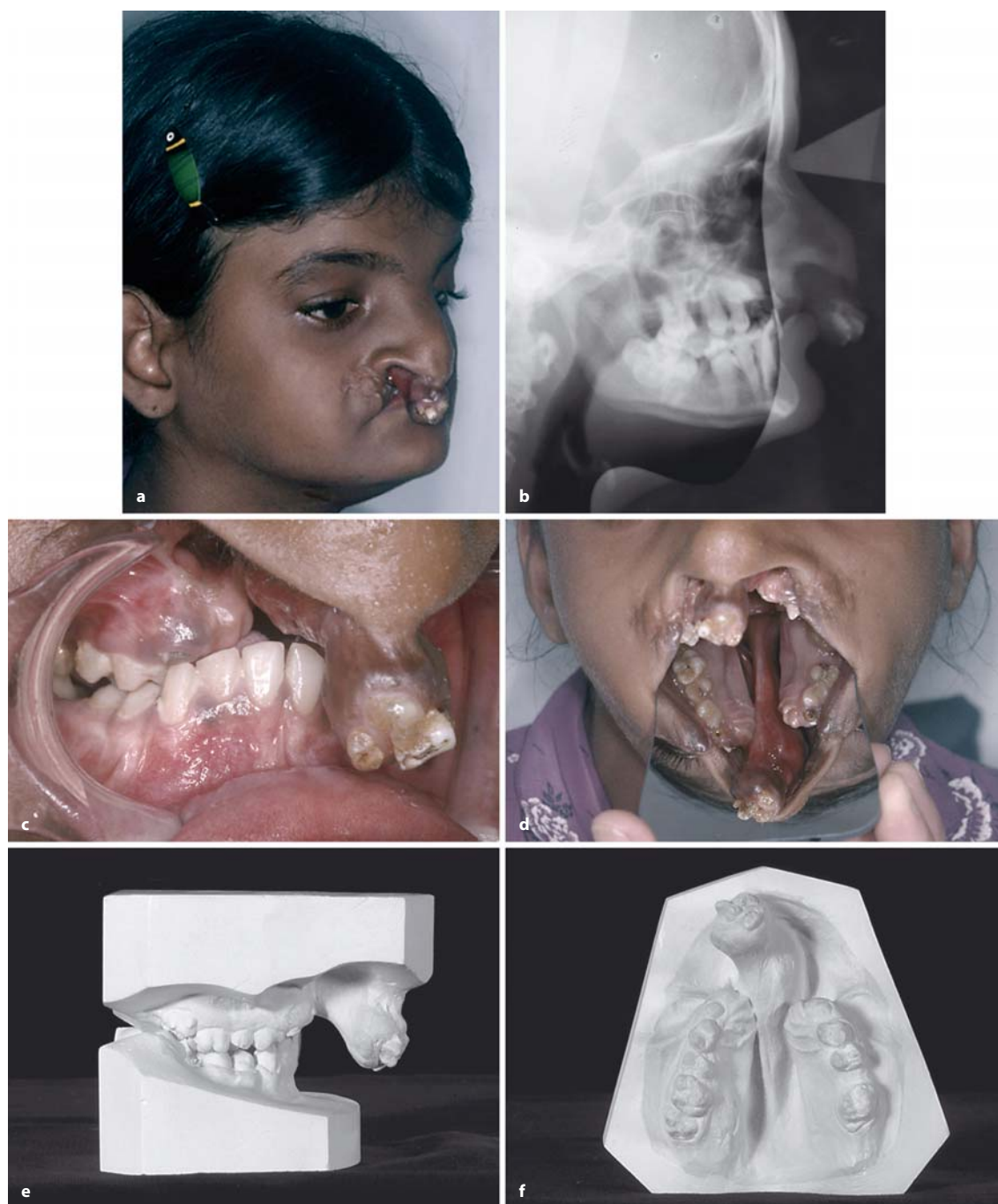
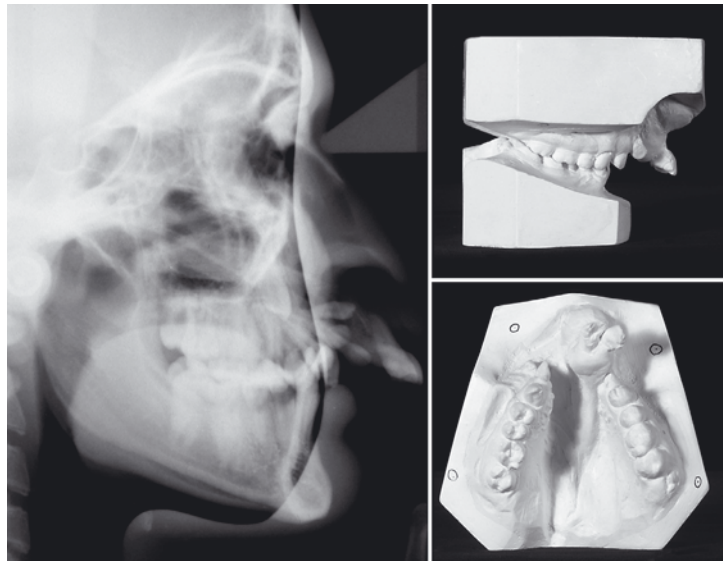


Fig. 10.11 a-f. Female unoperated BCLP unrestrained forward and downward growth of the premaxilla

Fig. 10.12. Male lateral skull x-ray and study models of unoperated BCLP. Note gross unrestrained forward and downwards growth of the premaxilla



10.4 Unoperated Isolated Clefts of the Palate

Unoperated isolated clefts of the palate present with relatively normal appearance. However, this group demonstrates more intrinsic deficiencies than clefts of the lip and palate. They have normal upper and lower dental arch relationships but with bimaxillary retrusion. They present with short maxillary length, small mandibles, and reduced upper anterior and posterior face heights. Figures 10.13–10.15 illustrate typical examples of the facial appearance and dental study models of unoperated isolated clefts of the palate.

10.4.1 Isolated Cleft Palate Study Models Analysis by Reflex Microscope [38]

10.4.1.1 Tooth Sizes

The tooth sizes in clefts of the secondary palate were found in general to be smaller than in the normal group, although these differences were not large and were clinically unimportant. The greatest differences were found in the incisor region. No differences were found between the sexes of the same group.

10.4.1.2 Chord Lengths and Perpendicular Distances

Chord lengths from the first molar to canine region were not different between the cleft and the control group but were different in the canine to central incisors distance. This reflects the consistently smaller size of the teeth in the patients with clefts of the secondary palate. In addition, perpendicular distances were generally smaller.

10.4.1.3 Arch Widths

The arch widths of the clefts of the secondary palate were narrower than the controls. This may be an adaptation of the dentition to the smaller and narrower mandible in this group.

10.4.1.4 Overjets

The overjets in the cleft group were slightly increased compared to the control despite the fact that the chord lengths and the perpendicular distances were shorter. This again may be a reflection of small mandibular size.

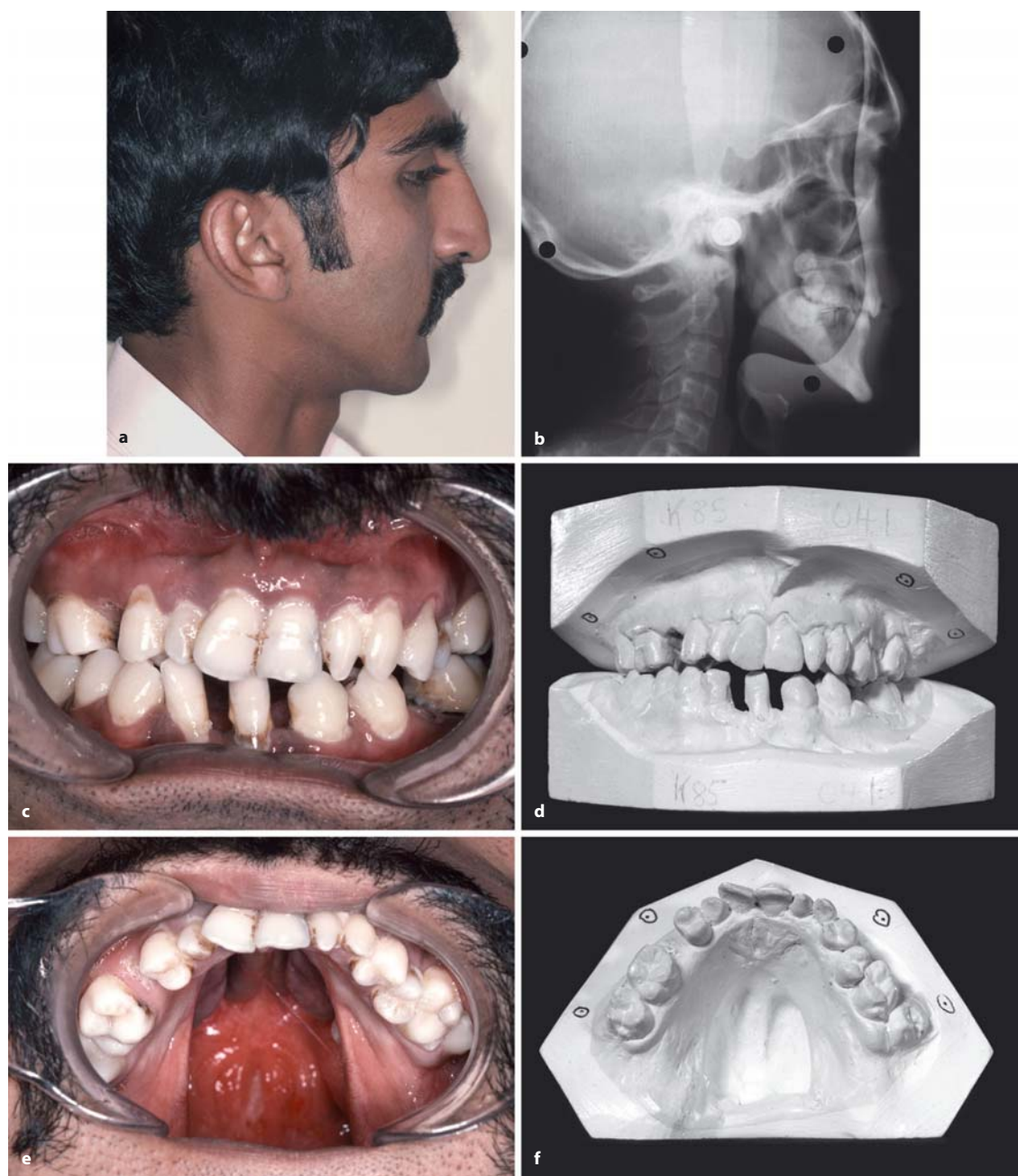


Fig. 10.13 a-f. Adult male subject with isolated cleft of the hard and soft palate



Fig. 10.14. Mature female with U-shaped ICP



Fig. 10.15. Mature female with V-shaped ICP

10.4.2 Factors Influencing Interpretation of Results from the Sri Lankan Cleft Lip and Palate Project

10.4.2.1 Malnutrition and Growth

It should be recognized that the above studies are derived from subjects in the developing world. Although Sri Lanka is a relatively advanced developing country, there is nevertheless significant malnutrition and endemic infections, for example, malaria.

The failure of infants with clefts to gain weight adequately has been documented by several authors [39, 40]. There is increasing evidence to suggest that poor nutrition in early life may be an important factor in growth disturbances seen in later life [41].

However, if there is failure to thrive or no catch-up growth by the age of 2 or in some papers by 5 years then perhaps attainment of normal limits for height, weight, or body mass index can never be expected.

Malnutrition, particularly at a period of especially rapid growth such as in utero, has long-lasting effects. Subsequent influences of undernutrition in the first and second years of life or later in childhood leave a long-lasting, complex growth problem. During childhood, stature is determined by the size that an infant has reached by the end of the first year of life, which is partly determined by genetic circumstances and influenced greatly by nutrition and the subsequent rate at which the child grows.

Nutritional status has a profound effect on growth hormone secretion. Malnutrition is a well-recognized form of reversible growth hormone resistance, which can be normalized with nutritional supplements. A malnourished mother is likely to give birth to a baby with low birth weight, while children with protein-energy malnutrition do not grow as well as others, according to a recent report [42]. This kind of malnutrition is an underlying cause of almost one third of the deaths among children under 5 years in Sri Lanka. Malnutrition is still a serious problem in Sri Lanka [43]. Food insecurity is one of the major reasons for malnutrition in Sri Lanka according to the Department of Census and Statistics. Poor financial and physical access to food is responsible for the malnutrition and food insecurity. Drastic price increases of essential food commodities and stagnating or deteriorating incomes created poor financial access to food. The civil war from 1984–2002 in Sri Lanka has exacerbated the essential food and financial problems.

A recent survey of 16,000 Sri Lankan children found that only one quarter were properly nourished [44]. More than one third were suffering from third degree malnutrition, the level beyond which children exhibit distended stomachs and skinny frames. Supporting evidence from the National Peace Council indicated that only 4,863 children under 5 years out of a random sample of 16,767 were within normal nutritional limits. Six thousand three hundred and seventy one children had third degree malnutrition, 3,186 with second degree malnutrition, and 2,347 with first degree malnutrition [45]. According to this report, diseases such as malaria, which is still prevalent in Sri Lanka are a primary cause of malnutrition. Worm infestations are the second most common cause. Lack of food itself is the third cause.

In another supporting paper it was found that Sri Lankans require a calculated average of 2,260 calories per day. Availability of protein has gradually increased; nevertheless a high incidence of malnutrition exists with 60% of children under 5 suffering from malnutrition [46, 47]. Poor growth of preschool children, high rates of low birth weight babies, poor maternal nutritional status, and micronutrient deficiencies are common nutritional problems in Sri Lanka.

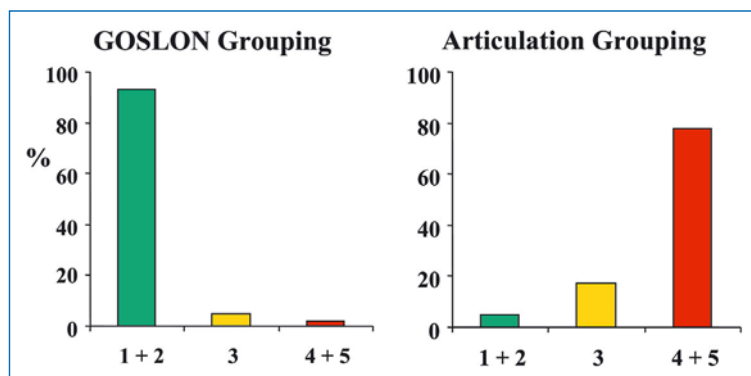


Fig. 10.16. Outcome for unoperated subjects beyond 13 years of age ($n = 42$). In both GOSLON and Articulation groupings 1 and 2 are excellent or good, while groups 4 and 5 are bad or awful

To measure quality of life in a nation, the United Nations Development Program [48] started figuring a Human Development Index (HDI). A nation's HDI is composed of life expectancy, adult literacy, and gross national product per capita.

There are vast differences when comparing or studying a different ethnic culture. The HDI for the UK is ranked as number 13 out of 130 nations. Sri Lanka is ranked at 79 – much lower than the UK. The comparisons between the two countries are shown in the table below.

UK and Sri Lanka Human Development Index

	UK	Sri Lanka
Life expectancy (years)	77.7	72.1
Total population (millions)	59.4	18.9
Annual population (growth rate)	0.1 %	0.8 %
Population under age 15	19 %	26.3 %
Undernourished people	0	23 %
Children underweight for age	0	33 %
Children underheight for age	0	17 %
Infants with low birth weight	8 %	17 %
Malaria cases (per 100,000 people)	0	1,111

Nutrition or subsequent malnutrition is only one environmental factor that can leave a long-lasting complex growth problem. Emotional deprivation also has a profound influence on the growth process and may interact with the provision of food [41]. A well-loved child is fed and nurtured, whereas a child with no prospect of a job or marriage or who is a burden may not be. Many of the subjects in this study were social outcasts, who dropped out of school. Females in particular were hidden away in their houses and only one female in the unoperated population married. Children need a good emotional climate to thrive. The mechanism of the effects of emotional deprivation on

growth is not well documented but is linked to reduced growth hormone secretion and its associated growth failure.

10.4.2.2 Speech Implications

While facial growth in the unoperated subject presents without maxillary retrusion, unlike many operated patients, the speech outcomes for the same series of patients demonstrate almost unintelligible speech for the whole sample. This is illustrated in Fig. 10.16, which compares and contrasts the GOSLON results with the speech articulation outcomes for a group of UCLP subjects who had both dental study model and speech recordings.

Research on the Sri Lankan Cleft Lip and Palate archive has demonstrated that surgery, when delayed beyond 8 years of age and even earlier, results in permanent irremediable speech disorders [17, 49].

10.4.2.3 Racial Variation

The Sri Lankan Cleft Lip and Palate project was careful to use healthy control subjects from the Sri Lankan population. Sri Lankans present with bimaxillary protrusion, females on a skeletal 2 dental base and males on a mild skeletal 3 dental base. These are reflected within the cleft population. Interestingly, only one subject presented with a class II/2 dental arch relationship on a skeletal 2 dental base (Fig. 10.17).

In a recent (2003) project in Gujarat and Rajasthan in northern India, the vast majority of patients presenting had class II/2 dental arch relationships on skeletal 2 dental bases. This further emphasizes the need to have control subjects from the same racial grouping (Fig. 10.18).

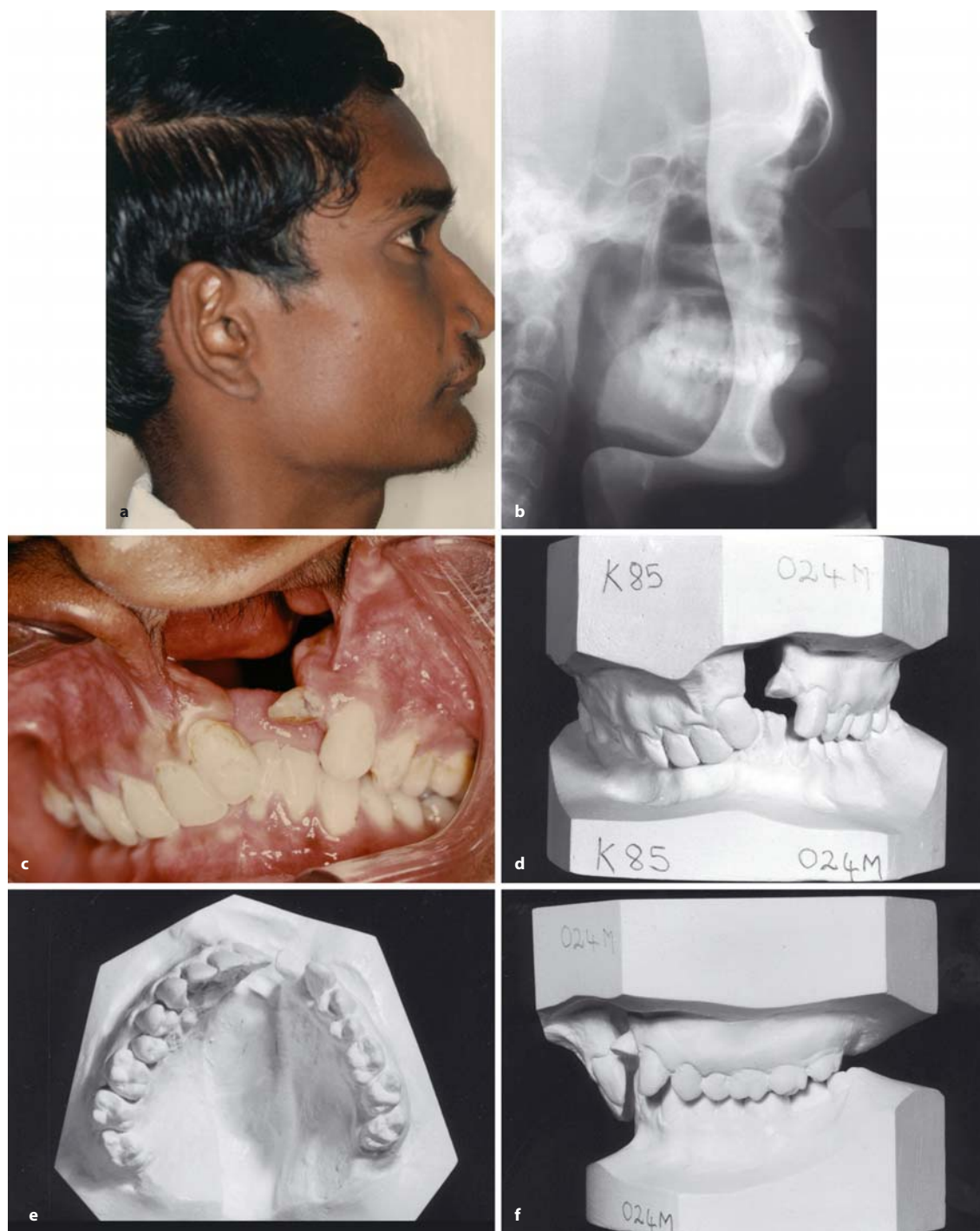


Fig. 10.17 a-f. Adult male subject with unoperated unilateral cleft of the lip and palate. This is a rare example of class II/2 arch relationship demonstrating retroclined upper incisors.

Very broad upper arch and very low FM angle. The lower lip controls the upper labial segment causing retroclination



Fig. 10.18 a, b. This occlusion and facial pattern is characteristic of patients in northern India

10.4.2.4 Surgical Implications

It should not be inferred that because unoperated cleft lip and palate subjects grow relatively normally, surgical regimens delaying palatal surgery are indicated. This study does not examine the effects of surgery or the timing of that surgery upon facial growth outcomes.

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11A A Brief Overview of Psychological Issues in Cleft Lip and Palate

KATHLEEN A. KAPP-SIMON

A review of the psychological issues for children with cleft lip and/or palate (CLP) must take into account the social, emotional, behavioral, and cognitive functioning of the children. There is a complex interaction among genetically determined traits such as temperament and intelligence and environmental factors such as parenting styles and social milieu. These factors interrelate to affect a child's self-concept, social acceptance, behavioral adjustment, and school success. These factors also affect a child's ability to cope with the stress of growing up with CLP. This chapter will provide a brief overview of these complex issues as they relate to (1) social and emotional functioning and (2) cognitive functioning and school achievement in individuals with CLP.

11A.1 Social and Emotional Adjustment

The social and emotional adjustment of a child with CLP must be viewed as a developmental process rather than a static state. Factors that have the potential of affecting this process will be connected to parental functioning, child functioning, and societal acceptance.

When a child is born with a birth defect such as CLP, parents experience multiple emotions that disrupt the equilibrium of the family. Shock, sadness, fear, grief, guilt, and anger are some of the emotional reactions that parents report [11, 54]. The first months of life can be extremely difficult for parents of a child with CLP as they handle their feelings, learn to feed their baby, and work to integrate their baby into the family. While most families are able to cope with this crisis and reorganize to meet the child's needs, some struggle [10]. Parental success during these first months of life will depend in part on the individual mental health of each parent, their coping skills, and the strength of the marital relationship. Parents who report high levels of stress during infancy that persists

into toddlerhood also report higher levels of adjustment problems for their children [39]. However, research also suggests that mothers who believe that they are able to meet their child's needs, who take satisfaction in their role as parents, and who are able to openly acknowledge the stress of the current situation are best able to nurture their infants in a manner that promotes healthy attachment during the early childhood, better emotional self-regulation during the preschool years, and fewer behavior problems during early elementary school [9, 52].

As the child grows, the role of the parent also develops. Parents serve as the child's first model of how to handle the difficult social situations that individuals with facial differences encounter. Parents also provide emotional support, impart positive discipline, and teach their child how to effectively relate to peers and adults. Children whose mothers reported less stress and greater parenting confidence during the preschool years were rated as more socially skilled, self-confident, and well adjusted when they entered the primary grades [24]. Similarly, Pope and Ward [40] found that parents who were anxious worriers about their children's social acceptance, but didn't act productively to assist their child, were more likely to have children who were having social difficulties while parents who actively encouraged their children's efforts to engage with peers had children who displayed higher levels of social competence.

Although parents have an important role in a child's long-term adjustment, child characteristics also have an effect on social and emotional functioning that is independent of the parent's behavior. Child development research has defined differences in temperament that influence personality types and are related to psychological adjustment. While there are a number of temperament systems, Hart, et al. [13] describe three broad personality types that provide a general guide to differences among children. These types are: (1) the resilient child who is socially compe-

tent, gets along with both adults and peers, tends to be gregarious, and exhibits positive emotions; (2) the overcontrolled child who typically exhibits extreme shyness, is quite compliant, and highly dependent on others; and (3) the undercontrolled child who is generally uncooperative, has difficulty with social relationships, is noncompliant, and more likely to exhibit negative emotions. These temperamental characteristics exist on a continuum and may be influenced by environmental factors.

Children respond to the stresses of growing up with CLP differentially based on their inborn temperamental characteristics. A resilient child tends to be more even tempered, tolerates surgeries with less distress, and is less likely to be bothered by questions about the scarring from his or her cleft. Because of their easy-going positive nature, these children are also less likely to be teased by their peers. If teasing does occur, they are more likely to view the teasing as the other person's problem. In contrast, a child who is overcontrolled and inhibited may demonstrate increased anxiety in social situations or when faced with surgery. This same child is more likely to feel stigmatized by his or her facial differences, believing that teasing from peers is an indication of a personal flaw. The child who is undercontrolled has difficulty complying with adult demands and may antagonize peers due to his or her inability to play cooperatively or comply with group mores. Thus differences in personality type may influence the way in which a child reacts to situations that are commonly encountered when growing up with CLP (see [16] for further discussion of how temperament affects coping).

Societal reactions also play an important role in the emotional adjustment of individuals with CLP. Multiple studies exist that demonstrate a relationship between facial configuration, attractiveness and the reactions and judgments that observers make about an individual, with more attractive individuals receiving positive attributions and less attractive individuals being judged more harshly [1, 7, 20, 25, 30, 32, 56]. Langlois et al. [26] completed a meta-analysis of studies on attractiveness and concluded that: "Beauty is more than just in the eye of the beholder; people do judge and treat others with whom they interact based on attractiveness; and perhaps most surprisingly, beauty is more than skin deep."

Thus it is not surprising that parents frequently attribute difficulties with peer teasing, exclusion from the peer group, or choosing to play with younger (safer) children to facial differences or accommodations of their children to those facial differences [17, 27, 38]. Objective ratings of facial difference have been associated with increased behavioral inhibition [47], which may also be related to quality of social interaction [19]. Focus group interviews with adolescents

who have facial differences concluded that these teens attribute their perceived lack of welcome by peers to their facial appearance [8]. A natural response to a perceived lack of welcome may be to limit social contacts to one or two peers with whom a child feels safe or the child may choose solitary activities. Either strategy may serve to decrease anxiety [14, 29, 43].

11A.2 Cognitive Development and School Achievement

The majority of children with CLP demonstrate intellectual development that places them within the broad range of normal [6, 12, 31, 50] although mean scores tend to be 3–5 points lower than the population mean of 100, particularly in the area of verbal skills [45]. A recent controlled study examining intellectual functioning of adult males with CLP compared to an age-, gender-, and SES-matched comparison group replicated the findings of intelligence research on children [37]. The mean full scale intelligence quotient (FSIQ) for male adults with CLP was within the normal range (FSIQ = 96.96, SD = 13.2) but 12.5 points below the control group (FSIQ = 109.5, SD = 9.27). Verbal intelligence was not more impaired than nonverbal intelligence for the adult subjects; however, verbal fluency was significantly less well developed for the subjects with CLP even when FSIQ was controlled.

The discrepancy between verbal and nonverbal intelligence has been researched in some detail by Richman and his colleagues [42, 44, 46]. The hypothesis of this body of research has been that children with CLP demonstrate two broad types of language-related deficit. One type involves a mild verbal, expressive deficit that has been associated with developmental reading problems and the other is a more pervasive language disability that affects language comprehension and associative reasoning abilities.

The incidence of learning disabilities, particularly reading disabilities, in children with CLP is significantly higher than the rate found in the general population, with an estimated rate of approximately 30%–40% [5, 46]. Recent research on the etiology of reading disabilities in children with CLP has identified several areas of interest. Continued research by Richman has identified deficits in rapid naming and automatic verbal memory, similar to the dysnomia model of dyslexia, as a primary deficit for children with CLP who have a reading disability [48]. Further investigation of this memory deficit suggests that children with CLP do not automatically provide a verbal label to information that is presented in a visual form even though their short-term memory for the same information presented orally is not impaired [49]. According to Richman, an important prelimi-

nary recommendation from this line of research is that children with CLP and reading disabilities would benefit from training in verbal labeling and oral phonics using a program similar to that recommended by the NICHD branch on dyslexia [28]. Despite articulation problems secondary to cleft palate, children with CLP should be taught using a phonetic approach rather than a whole word or sight reading approach.

Reading and language disorders in children without CLP have been associated with changes in neuroanatomy and function [21, 22, 41, 51]. Nopoulos and her colleagues have postulated a similar relationship between CLP and brain structures and function [37]. The focus of Nopoulos' research to date has been adult males with CLP. She has found changes in brain structure including enlargement of the anterior regions of the cerebrum, with a corollary decrease in size of the lateral and posterior regions of the cerebrum and a decrease in size of the cerebellum that have been associated with cognitive functioning; the greater the change in brain structure, the lower the FSIQ [34–36].

A significant number of children with CLP are considered developmentally “at risk” during infancy and the preschool years [18, 23, 33, 54, 55, 57]. An “at risk” classification generally indicates that the child is not progressing at the expected rate; however, the long-term implications of that delay are not always clear. In the case of children with CLP, developmental delays may be considered secondary to known biological risk in the form of early disruption of cranial cell migration and, therefore, indicators of possible learning difficulties during the school years (see Aylward's work for a discussion of neuropsychological risk in infancy [2, 3]). Since early intervention for developmental reading disabilities has been demonstrated to remediate the problems for a significant number of children, it is critical that Cleft Teams provide the necessary screening for cognitive deficits with recommendations to the families for early intervention services [4].

11A.3 Summary

Psychological factors including parental adjustment, child temperament, social and emotional adjustment, intellectual development, and school achievement for children with CLP were discussed in this chapter. The emotional, behavioral, and social adjustment of children with CLP is dependent on multiple factors. Some of these have been described in this short overview. It is clear that parents play a vital role in supporting their children with CLP and parental confidence and skill can facilitate good adjustment [9, 15, 39]. Nonetheless, characteristics intrinsic to the child including temperament and intelligence will also

make critical contributions to the child's overall well-being. While the interplay between intrinsic characteristics and environmental factors is not unique to children with CLP, recognition of these factors by members of the Cleft Team will assist them in understanding the needs of both children with CLP and their parents.

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11B Craniofacial Psychology: New Directions

JOYCE M. TOBIASEN

Francis Macgregor [1] described the plight of facially impaired patients over 50 years ago: “staring, remarks, curiosity, questioning, pity, rejection, ridicule, whispering, nicknames, and discrimination.” Macgregor’s [1] descriptions are consistent with social scientist’s descriptions of facial deformity as a stigmatizing condition – much like obesity, physical disability, mental retardation, homosexuality, and gender. A stigmatized group is defined as a “social category about which others hold negative attitudes, stereotypes, and beliefs, and which, on the average, receive disproportionately lower interpersonal outcomes relative to members of the society at large because of discrimination against members of the social category” [2].

There is no doubt, that people hold negative stereotypes of numerous groups including blacks [3], women [4], unattractive individuals [5], facially disfigured individuals [6], homosexuals [7], and mentally ill persons [8]. Moreover, these groups are clearly disadvantaged with regard to economics and barriers to advances in our society [2]. Individuals with facial deformities suffer numerous disadvantages including lower income on the average [9] compared to normal siblings. Individuals with clefts tend to marry later and less frequently on the average than their normal siblings [10, 11]. They reported experiencing problems in relationships with the opposite sex [12], and were teased more often by peers [13]. Harper and Peterson [14] conducted a series of studies worldwide that examined normal children’s attitudes toward children with obesity, orthopedic-mobility problems, amputation, and facial clefts. The body of research represents 25 years of study in five countries. Fourteen hundred children indicated consistent negative attitudes toward children with clefts over the other disability groups. Children with facial clefts were the least likely to be chosen as a playmate than the other

groups. Other studies found strong prejudice against individuals with clefts [15–17].

There has also been research on the content of the negative stereotypes. Weinberg and Santana [18] sampled comic books for cultural stereotypes of good and evil based on physical appearance. Individuals with irregularities from the neck up (i.e., distorted heads or facial features, scars and excessive wrinkles represented evil characters 71 % of the time, were never depicted in neutral roles and were represented as “good” only 29 % of the time. Tobiasen [19] examined personality and ability judgments associated by peers with cleft-related impairments. Children were shown either photographically corrected versions of children with congenital facial clefts or uncorrected versions. Children and adolescents, males and females, ages 7–16 years rated individuals with cleft impairments as less popular, less smart, and a less likely choice as a friend. Girls with clefts tended to be rated more harshly than boys.

The evidence that prejudice and discrimination have severe negative social, economic, and political consequences for stigmatized groups, in general, and facially impaired individuals in particular is incontrovertible. We will review the research on self-esteem for stigmatized groups in general and facial deformity in particular. The question of how individuals with facial deformities maintain normal self-esteem in an environment of prejudice will be explored. It is argued that individuals with facial deformities develop specific coping strategies to protect self-esteem. Finally, the implications of these coping strategies for research and practice will be discussed. The conceptual framework for this chapter is taken from Crocker and Major [2]. They documented how self-esteem in negatively stereotyped groups is protected by coping strategies that develop naturally in stigmatized groups.

11B.1 Stigma, Self Concept, and Social Psychological Adjustment

Social scientists have developed theory and conducted research on the relationships between stereotypes and expectancies toward given groups and self-concept for decades. Developmental [20, 21] and rehabilitation psychologists [22] have proposed that there is a multicausal chain between expectancies, behavior, and personality development. The theory is based on the concept of “self-fulfilling” prophecy [23], which asserts that people’s expectations influence the behavior and personality of others such that the expectations, behavior, and personality development are congruent. The interaction between the expectations of people for the behavior of others can set up a closed social interaction feedback system such that expectations influence behavior that feeds back to confirm the expectations.

Darley and Fazio [24] described a six component model of the social interaction sequence underlying the concept of self-fulfilling prophecy. First, a perceiver has expectancies toward a target person. These expectancies are based on past observations of an individual or on stereotypes that the perceiver may hold about specific categories of people. Second, the perceiver’s expectancies influence his or her behavior toward the target person. Third, the target person interprets the perceiver’s behavior. Fourth, the target person’s behavior reflects his or her interpretation of the perceiver’s behavior. Fifth, the perceiver interprets the target’s behavior as congruent with expectations. Finally, the expectations and behavior may become central to the target person’s self-esteem. The concept of self-fulfilling prophecy has been applied to explain the effects of teacher expectations on student achievement [25], sex differences in behavior [26], racial stereotypes, and the effects of the physical attractiveness stereotype [27].

If the expectancy-confirmation model of the social interaction sequence is applied to an analysis of facial impairment research, then we should be able to identify: specific expectations, differential behavior, unique interpretations by affected individuals of the behavior of others toward them, identifiable behavioral patterns associated with affected individuals, and self-esteem that has been negatively molded by the fact that an individual has a facial impairment. The focus of this paper is on self-esteem. There are several review articles that address overall psychological adjustment [28]. These will not be reviewed here. The assumption that individuals with stigmatizing conditions must have psychological problems has not been supported in the literature. Stigmatized groups do not consistently present with psychopathology and

low self-esteem. The lack of consistent findings for psychopathology led Clifford to observe “Why are they so normal?” [29].

The self-esteem of several stigmatized groups has been carefully studied. Self-esteem is defined as a generalized feeling of self-acceptance, goodness, worthiness, and self-respect. It is widely recognized as an essential part of healthy psychological functioning and is strongly related to general satisfaction with life. Specific self-esteem is differentiated from global self-esteem. For example, one may feel confident in a specific ability and yet feel unworthy in a general sense and vice versa.

Blacks have levels of self-esteem equal to or higher than that of whites [30]. Similarly, women on the average have comparable self-esteem to men. [31] Children described as unattractive have normal self-esteem [32] as do disabled individuals including individuals with mental retardation [33]. There have been at least ten studies of the self-esteem of children and adolescents with cleft deformities [34–43]. Two found lower self-esteem for the affected group, and eight found either normal or high normal self-esteem. Children with facial impairments do not differ on reported global self-esteem from normal groups. Overall adjustment is within the normal range with the exception of a pattern of reticent social behavior. The question is how do they maintain self-esteem and cope with societal stigmatization?

11B.2 Self-Protective Properties of Social Stigma

Crocker and Major [2] describe mechanisms that may buffer the self-esteem of stigmatized groups. These mechanisms are unique to stigmatized groups because they emerge from the experience of being in the group. First, Crocker and Major [2] describe a discounting mechanism, or attributing negative feedback to one’s group and not one’s self. For example, a woman may discount her lower salary compared to a man by attributing it to “being a woman,” rather than to her incompetence. Hence, self-esteem is protected. Second, stigmatized groups tend to compare themselves to similar groups on the stigmatizing characteristic rather than the more advantaged nonstigmatized group. For example, an individual who is intellectually challenged will be less likely to compare his school performance with gifted children than with others who are similarly challenged. Individuals with facial deformities may not compare their facial appearance to nonaffected individuals but to their own group. Third, stigmatized individuals may develop different values than the nonaffected group. They may devalue or minimize the importance of the stigmatizing char-

acteristic and value areas in which their group may be strong. For example, an individual with a facial deformity may value the characteristic of coping with a long surgical course of repair and hold less value in the importance of facial appearance as a defining personal characteristic.

This approach is related to an approach recently described by Eiserman [44] who asserts that studies of adjustment to facial deformity need to focus more on healthy adaptation and less on pathology. He argues that individuals should be considered on a continuum of health and disease. Such a continuum allows us to focus on a comprehensive view of an individual rather than on a simple etiological examination of deficits, i.e., facial deformity causes low self-esteem. Strauss [45] also raises the question of focusing on healthy behavior. "Why is it that, despite the numerous challenges and problems shown in the literature, many persons with cleft lip and palate and craniofacial conditions become productive, contributing, happy, satisfied individuals?"

11B.3 Research on Self-Protective Properties of Facial Deformity

There has been almost no specific research on whether or not individuals with craniofacial deformities use self-protective strategies to protect self-esteem. However, there are hints from various studies and observational data that lead to speculation that they do use these strategies. We will examine the three protective strategies described above, i.e., discounting or attributing negative feedback to the stigmatized group and not to the self; selectively comparing outcomes with members of groups similar to oneself; selectively devaluing the importance of facial deformity and valuing the attributes on which the group fares well.

11B.3.1 Attributing Negative Feedback to Prejudice

There is some evidence that individuals with facial deformities do attribute negative feedback to their group rather than to themselves personally. Individuals with facial impairments tend to assume that their facial appearance will result in negative consequences as a routine part of life. Abel [46] reported that all of her sample of 74 patients hospitalized for corrective surgery of facial impairments ranging from clefts to severe acquired impairments stated that their facial problems often resulted in social and vocational discrimination. Clifford [47] interviewed adolescents with clefts about the impact of the cleft on family life.

These children reported that they thought that their parents were unhappy with their facial impairments, but that they, nevertheless, were nurtured and accepted by their parents.

Several investigators [48–52] have observed that individuals with physical disabilities often take it for granted that the impairment has an influence on their social relations. Kleck and Strenta [51] conducted a series of four laboratory studies with nondisabled and nonimpaired college students that directly examined if individuals with stigmatizing physical characteristics expect that their treatment by others is causally linked to these attributes. The experimental paradigm involved a face-to-face interaction in which college students were given a mental set that involved 1 of 3 conditions; a facial scar, epilepsy, and allergy. The mental set for the scar was implemented with makeup that was removed without the subjects awareness prior to an interaction with a confederate trained to act in a standard manner and blind to the mental set given the subjects. Participants in the facial scar and epilepsy conditions believed that the behavior of the confederates that they interacted with was affected by their presumed impairment while those in the allergy condition did not. The subjects believed that the behavior of the confederates was influenced by the scar even though the scar, unbeknown to the subject, had been removed.

Strenta and Kleck [52] conducted a follow-up study in which individuals were given the same three sets described above and then were asked to perform a perceptual segmentation task that required them to mark off each time that they observed a "meaningful action" on the tape. Raters marked off significantly more actions when they were in a stigmatized group. This suggests that stigmatized groups may pay closer attention to the content of their interactions with non-stigmatized groups. They may be looking for discriminatory actions so that they can better prepare themselves and protect self-esteem.

Jensen [53] found that students who had been insulted because of their race, religion, nationality, or residence who attributed such insults to prejudice against their group were equivalent in self-esteem to students who had not been insulted. There have been studies of the self-protective function of attributing negative outcomes to prejudice in which women or members of minority groups received negative feedback or poor outcomes from an evaluator who might be prejudiced against them. In one study, female subjects received negative feedback from a male evaluator. The subjects who believed that they had been discriminated against had higher self-esteem [54].

There are no direct studies of the attributions facially deformed individuals make to negative social feedback. However, if they attribute the negative feed-

back to their group rather than to their person, self-esteem is protected. Crocker and Major [2] speculate that, if positive feedback is given to the stigmatized group, then one may be more likely to attribute it to one's real ability, or to pity.

Finally, there is a report that attributions to the stigmatized group can be overused and result in a pathological adjustment. Baker and Smith [55] described an extreme case of using prejudice against the group to explain all negative outcomes in life. "For years, the scar, harelip or misshapen nose has been looked on as a handicap and its importance in the social and emotional adjustment is unconsciously all embracing. It is the hook on which the patient has hung all inadequacies, all dissatisfactions, all procrastinations and all unpleasant duties of social life and he has come to depend on it not only as a reasonable escape from comparison, but as a protection from responsibility."

11B.3.2 Selective Comparisons to Similar Groups

According to social comparison theory, there is a well-documented tendency for us to compare ourselves to those who are similar on a given dimension. Researchers found that people prefer to compare their abilities with others who are similar to themselves [56]. Researchers have shown that this extends to comparisons of both outcomes and abilities [57]. Stigmatized individuals may avoid comparisons of facial appearance relevant dimensions to nonstigmatized groups because the comparison is too painful. It may be more adaptive to focus on dimensions in which the individual or group do well.

Eiserman [44] conducted a combined survey and interview study of 22 individuals with craniofacial deformities. There is some evidence that individuals with craniofacial deformities do see themselves as a unique group with special strengths and weaknesses. The focus of the study was to better understand the potential ways a facial deformity may have a positive impact on affected individuals. One respondent described how as a child he was an observer, rather than a participant with peers. He did not identify with his peers. They were in their group and he was in a group of one. "I think I was on my toes as a kid, paying very close attention to what was going on around me. If someone was going to make fun of me, I wanted to see it coming so I could brace myself or prepare to respond." This comment suggests that not only was this individual vigilant when interacting with advantaged others, but careful about discriminating the others as a group separate from him, hence minimizing social comparisons with that group.

Eiserman [44] asked his respondents: "Given the way you perceive your experience with facial differences to have affected your life, if you could live your life again would you choose to completely remove this experience of facial difference from your life?"

The findings indicated that more than one half of the affected adults and slightly less than half of parents would not choose to remove the experience. The responses indicated pride in being part of their group. An individual who would choose to remove it also responded positively: "Having a cleft lip and palate has helped create much beauty inside of me and I definitely honor what I've become because of it." Another individual who would not remove the deformity responded: "While I would give up the hard part in a second, I have a feeling that I would give up a central part of who I am that makes me the person I am if I sacrificed the experience of having a facial difference." The findings from this pilot study suggest that individuals with facial deformities compare themselves to their facial group. They view the members of the group as heroic and beautiful.

11B.3.3 Selectivity of Values

Crocker and Major [2] suggested that stigmatized groups may protect self-esteem from negative feedback or negative comparisons by others by selectively minimizing the importance of dimensions on which their group may fare poorly and selectively valuing those dimensions on which their group does well.

There is experimental support for this view in the social psychological literature. Harter [58] reported that children with high self-esteem tended to regard the areas in which they excelled as more important than those in which they did less well.

There is some evidence that adolescents with cleft deformities use different values to evaluate the severity of a facial difference than to nonaffected individuals. They tend to minimize the perceived severity of cleft deformity. Tobiasen and Hiebert [28] reported on a series of studies that addressed the following questions: what are the relationships between self, peer, and parent ratings of severity of cleft impairment; what are the relationships between these ratings and measures of self-esteem? A standardized measure of severity of facial cleft deformity was used. It was argued that individuals with severe facial clefts and their parents would minimize the severity of the impairment in comparison to nonaffected peers. Moreover, the minimization of severity would be associated with higher self-esteem and better social adjustment. Correlational analyses were conducted to examine the relationships between self, parent, and

peer ratings on the Facial Impairment Scales for Children (FISC) [59]. The cleft group FISC ratings were not correlated to peer ratings ($r = .30$). Cleft group FISC ratings of the self and parent ratings were significantly correlated ($r = .63$; $p = .006$). About 32 % of cleft patients rated themselves as significantly less impaired compared to their peer ratings.

Subsequent analyses were conducted to evaluate the relationship between the FISC ratings and the measures of adjustment. Patients who rated their impairment as less severe relative to peer ratings had significantly more positive ratings of global self-esteem, social competence, and psychological adjustment compared to patients who rated their impairments as equal to or more severe than their peer ratings. Interestingly, children with the least severe impairments reported lower self-esteem and more adjustment problems than those with the most severe impairments. Children who minimized the severity of the facial impairment had higher self-esteem and social competence, and better adjustment. The more severe the deformity, the better the adjustment scores and the higher the global self-esteem ratings. The findings suggest that an individual's self-perception of facial deformity may be a better indicator of good psychological adjustment than peer ratings. These findings give preliminary support that adolescents with cleft deformities use a different yardstick to measure severity of cleft deformity.

There is also clinical evidence that adults with facial deformity develop positive values about their lives that may be unique. Eiserman's [44] study participants reported that they had special attributes that developed in the course of coping with facial impairment. Several study participants reported that they had unique or enhanced communication skills: "It wasn't enough to have average social skills. I needed unusually good skills to deal with all the strange things other people did when communicating with me." Another participant reported "part of how my personality seemed to have interacted with having a facial difference was that I had to compensate for how I looked so that people looked beneath my face."

A second value was commitment to service. "My experiences of discrimination based on appearance have made me more aware of social justice issues and motivates me to do what I can to make the world a better place to live in." Third, several participants reported that they had acute observation skills: "I am acutely aware of things and am able to understand many subtle things about many social situations...". Fourth, study participants described themselves as more tenacious than others: "I've been increasingly described as tenacious as the years go by...". Fifth, an orientation to question society's beliefs was described: "I see so many people who truly are disfig-

ured by taking on society's distorted view of the importance of beauty." Sixth, participants described how they had a diverse social circle. "I have friends who are from diverse cultural backgrounds, one who is a burn survivor, gay and lesbian friends... and I think that my own experience of difference helped me to be open to people I most likely would not have been open to..." Finally, study participants described themselves as "normal". "I don't like the idea of even being asked if there is something extra positive that comes from an experience like having a facial difference. This is just like any other experience life has in store for us. There isn't a single person alive who doesn't have incredible difficulties at times, including disappointments, loss, failures, rejections, etc." These comments excerpted from Eiserman's [44] thoughtful paper do not fit the stereotype of the victim of stigmatization. Rather, these individuals coped extremely well and they attributed a strong commitment to unique positive values as essential to their success.

In this section, we examined three properties of stigmatized groups that may contribute to overall positive adjustment, high self-esteem, and normal lives. Much research is needed. What we know is speculative. However, social psychologists have studied self-protective properties in other stigmatized groups and the evidence is clearly supportive. Hence, this is an inviting new direction for study in craniofacial psychology.

11B.4 Surgery and Self-Esteem

Surgical intervention is of course available to treat facial deformity and to potentially improve evaluation by others and self-esteem. Removal or minimization of the facial deformity ought to reduce stigmatization and possibly improve self-esteem. However the relationship between surgery and self-esteem is complicated.

Behavioral scientists have traditionally been in favor of early intervention. Early intervention may reduce stigmatization and foster healthy development. Clifford [47] argued for early intervention because the longer an individual has to cope with a craniofacial deformity, the greater the dependence on it and the greater the problem when it is removed. However, the length of time of living with a facial deformity may be necessary to the development of self-protective strategies. If an individual has an early intervention that either removes or minimizes the deformity, and then later suffers a developmental malformation, how is self-esteem affected? The developmental malformation has the character of an acquired deformity. Individuals with acquired deformities may not have developed self-protective strategies and may suffer

self-esteem problems. It may take time for them to develop those strategies. In order to cope with a stigmatizing condition, one must be in a stigmatized group. If an individual has no experience in the group, then they may not be prepared to cope with the stigma. These issues raise many new and interesting questions about the timing of surgical intervention. How are self-protective strategies affected by surgery? Team psychologists need to address these questions with patients and educate them about what they can do to better adjust to surgical change in their faces.

The study of the relationship between surgery for facial deformity and self-esteem has produced confusing findings. Lefebvre [60] reported the results of a study of 225 children with Apert's syndrome. Parents' ratings of the children's appearance improved significantly 1 year after surgery. Self-esteem of the children was also significantly higher 1 year after surgery. However, 4 years later, self-esteem returned to the presurgical level. Surgery was not a lasting boost to self-esteem. Kiyak [61] found that self-esteem improved in anticipation of surgery to correct a facial deformity, but decreased after surgery and 24 months later. Other researchers reported on the beneficial effects of surgery and positive psychological adjustment [62, 63]. However, these investigators did not report long-term follow-up. Kapp-Simon [64] wrote that individuals who still have facial deformities after surgery have different adjustment issues. The residual deformity may still be a stigmatizing condition. If individuals believe that they will no longer belong to a stigmatized group following surgery, they may be disappointed. If they are carefully prepared and informed of a possible residual deformity, then they will be better prepared to deal with stigmatization. Kapp-Simon [64] reported that some patients regretted surgery because they still felt stigmatized. Patients need help to cope with surgery and its aftereffects. Education about coping skills is also needed. Clinical psychologists can play an important role in developing standardized educational materials to assist patients.

developmental malformations? Do they lack experience in coping with stigma and are therefore more vulnerable to attacks on their self-esteem? Can these techniques be taught? When is the best time to do that?

Research on whether or not individuals with facial deformities use self-protective strategies needs to be done. We already know that they do make attributions for negative outcomes to prejudice. However, we do not know how widespread this is. More importantly, we do not know if the external attribution serves a protective function for self-esteem. This question could be handled using an experimental paradigm with either or both children or adults.

Little is known about the social comparison processes of individuals with facial deformity. Do they selectively compare themselves to normal peers? Do they compare themselves with peers on sports, academic achievement, and other abilities but not on facial appearance? Who do they compare themselves with on facial appearance, or is facial appearance a less relevant attribute than other dimensions? After years of attending craniofacial clinics, do other clinic patients serve as a reference group?

Finally, further study of the values of individuals with facial deformity needs to be done. Does the facial deformity group use a different yardstick for evaluating facial deformity so as to minimize its severity? How and when do unique group values develop?

These are ambitious questions and they lend themselves to multi-institution involvement in order to have the needed numbers of patients so that controls for the many variables that may influence adjustment can be made. These variables include: age, diagnosis, severity of deformity, hearing problems, speech problems, level of intellectual functioning, and socio-economic status. It is exciting to consider a new direction in social psychological research on facial deformity. Affected individuals are clearly not passive victims of stigmatization, but active, adaptive human beings. Like all of us, they belong to many groups with no one group providing a total identity.

11B.5 Research and Clinical Implications

Recognition that individuals with facial clefts may develop effective resources to cope with social rejection opens up a variety of research questions not previously addressed. Little is known about the development of protective strategies. This question is extremely important, especially for clinical application. When and how do children start to use coping strategies? How do parent attitudes toward the deformity influence the development of protective strategies? How is change in the deformity accommodated? For example, how do individuals cope with residual deformities and de-

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Clefts of the lip have probably been with us for ages. Clay models of people with clefts in Peru apparently date back as much as 3,000 years. However, there is no mention of clefts in the writings of Hippocrates [1]. (460–375 BC), Celsus (25 BC–50 AD) or in the Edwin Smith papyrus [2, 3]. In China, there is a written record according to Morse as follows, “In the Chin Dynasty (229–317 AD), there was a surgeon who did plastic surgery for harelip. In the Tang Dynasty (618–901 AD) another surgeon, Fang Kan, was designated as “the doctor of lip repair” from which he obtained considerable repute [4]. The Indian writings of Susrata also refer to lip surgery, but in western Europe, it would be many years before lip repairs were attempted on a regular basis. The first written reports appear in the Middle Ages with the description of a cleft lip repair by Jehan Yperman (1295–1351 AD) a Flemish surgeon [87].

Clefts of the lip or the prepalatal structures (the primary palate) include not only the lip, but also the nasal tip, the alveolus, and anterior hard palate back to the incisive foramen. The defects vary considerably as to their extent, whether the cleft goes “completely” through these structures or only partly through (“incomplete”) and to how much the involved structures are displaced or underdeveloped, also whether the defect is unilateral or bilateral [5]. Prepalatal clefts are usually associated with palatal clefts and may be associated with other facial clefts or bodily anomalies.

Ruyao Song, a student of Dr. Robert H. Ivy and an outstanding plastic surgeon in China, noted in his preface to a publication of cleft lip repair [5] that we should draw a distinction between a “concept” and a “technique.”

Dr. Song wrote, “A concept is an idea, and a technique is the method used to carry out the idea. The technique may involve artistic planning, physiological considerations, and the operation itself. Thirty-seven hundred years ago, the Koomas of ancient India had the concept

of transplanting a pedicle flap from the forehead to reconstruct the lost nose. As a result, the technique of total nose reconstruction was originated. Four hundred years ago, Branca and Taglicocci of Italy had the concept of using a pedicle flap from the upper arm to reconstruct a new nose; hence, the technique of transplanting tissue from a distant part of the body was initiated. In the late 1950s, a plastic surgeon had the idea of making the forehead flap an arterial island flap and transplanting the neurovascular pedicle subcutaneously; hence, the method of total nose reconstruction in one stage came into use.

Although countless examples like these can still be cited, there is no doubt that concept is the mother of technique. However, knowledge begins with practice. Most new ideas emerge after repeated and diligent practice of the method. Again, the technique is the cradle of concept.

In prepalatal clefts, many techniques have arisen to repair the unilateral and bilateral cleft of the lip and to reconstruct the nasal deformity from the asymmetry of the unilateral cleft lip nose to the more symmetrical, but distorted bilateral cleft lip nose. Repair of clefts of the lip itself have developed from just trying to get the two edges together and to heal, to achieve symmetry, reconstruction of the Cupid’s bow and the development of systems of measurement to give reproducibility. To be able to do this reliably one must recognize that the Cupid’s bow on the cleft side is usually present on the medial side of the cleft [6]. The remnant simply needs to be brought down to the horizontal position. Then there are operations to reconstruct the distorted orbicularis muscle [7].

Repairs of the nose were originally just closure of the nasal floor. Then came undermining of the tip cartilages, then undermining and repositioning these cartilages and flap procedures to relieve web contractions in the nasal lining. The reconstruction of the deficient columella in the bilateral cleft has similarly

evolved from using tissue in the philtrum, the nasal floor or composite grafts from the ear to repositioning the tip cartilages and more recently stretching these tissues preoperatively with an apparatus attached to a dental plate [8].

The problems with the alveolar structures have led to even more controversy and divergences of opinion. This has been complicated because unlike the repair of the cleft lip where one can see how close to symmetry one has come and where there is little change with time (i.e., if the Cupid's bow on the repaired side is higher than on the normal side at the end of the operation it is likely to remain so over the years) [9], the cleft alveolus is usually not only lacking bulk and distorted in position, it also has misshaped, malpositioned, or absent tooth buds and its growth can be severely limited by scars. Even with careful surgery, there is concern about inhibition of subsequent growth. In addition, in the bilateral complete cleft, the prolabium of the lip does not contain innervated muscle. If the muscle is not reconstructed across the midline at the time of cleft lip repair, the lateral lip structures will usually stretch or "expand" the midline skin and mucosa, leaving the central portion of the lip conspicuously wide.

There have been many opinions as to when is the best time to repair the cleft lip. Some say that the newborn age is the best because one may be able to garner some of the benefits of "scarless" fetal healing [10]; however, most results of these authors do not seem to show this convincingly and it is unlikely that the newborn will benefit from "scarless" fetal healing. Early repair does give the parents a much more acceptable-looking child, which should be considered in the decision on timing.

Most surgeons presently prefer to perform the lip repair at about 3–4 months of age. At this time, the child will have passed the usual neonatal drop in hemoglobin, and be able to manufacture red and white blood cells fairly well, which can be important for anesthesia and for warding off infection [11]. Further, the older child will have better-developed lung structures, respiratory physiology, and responses to anoxia. In addition, these children have a higher-than-normal incidence of other anomalies and these might not be apparent in the newborn period.

There are studies that show that the adult with an unrepaired cleft has more normal facial bone growth than the child with a cleft which has been repaired in infancy or early childhood. We stress the word "more" because Dr. Fernando Ortiz Monastario, who has studied a number of adults with unrepaired clefts, is usually *misquoted* as saying that these adults have "normal" facial bone growth. He really did not say this and we have never seen an adult with an unrepaired cleft who has "normal" facial bone growth – except for

the most minimal clefts (or form fruste), which usually have fairly normal facial bone growth. He did say that they had *growth that is more normal* and that the "angle of convexity" on profile was often greater than normal [12]. However, every complete cleft that we have seen – unrepaired in an adult – still had considerable alveolar deficiency at the alveolar cleft with significant vertical displacement, and often-appreciable collapse of the maxillary lesser segment. Monastario has agreed with these statements.

The timing and extent of surgery on the nasal tip has also been subject to much controversy. At one time, outstanding rhinoplasty surgeons who handled mostly adults (Gustaf Aufrecht, for example) were pleading, "please do not touch the nose of a child with a cleft before the child is fully developed. You produce so much scar and leave so much deficiency that it is almost impossible to correct the deformity in the adult patient" [13]. Fortunately, the techniques of cleft lip nasal tip repair in the early age group must have improved because we seldom see that problem today, though most children with severe clefts who have surgery to the nasal tip in infancy are very likely to need some further "touch up" surgery when they are older.

A Word on Technique. The earliest repair of cleft lip usually consisted of incising or "freshening" the cleft edges and sewing the raw tissue together. There were many hazards to this surgery. For the most part, it was done very quickly and without anesthesia. The tissues are very vascular and control of bleeding was not good. Further, with the lack of aseptic surgical techniques, sutures in and about the mouth frequently became infected. For cleft lip repairs, often closure was achieved by transfixing the tissues with silver needles that had sharp steel tips and then wrapping linen



Fig. 12.1. A steel-tipped silver needle was used in this fashion in 1676 to hold the lip incision together "winding the thread about, as Taylors do when they stick them on their skirts." (With permission from [14])

thread around the protruding ends of the needles in a figure-eight pattern [14]. It was felt that sutures in the gingival or the palatal tissues had to be removed by the end of a week to avoid serious infection (Fig. 12.1).

The first lip repairs were simple straight-line closures, which left the shape considerably distorted. Blair, Brown, Byars, and McDowell introduced the design of a flap just above the vermilion and a system of measurements to achieve reasonable symmetry. For years – at least in the USA – this was the preferred technique for cleft lip repair [15]. Though these lips were often symmetrical, the technique ignored the Cupid's bow and usually had a high point in the center. Many different flaps and designs have been advocated. Mirault in France described a flap similar to Blair's, so the St. Louis surgeons added the name of Mirault to their manuscript. Hagedorn in Germany described a square-shaped flap and LeMesurier in Toronto described using such a flap to cause a bend in the vermilion to construct the peak of the Cupid's bow on the repaired side [16]. Tennison described a fairly large triangular flap in the lower one third of the lip with very nice results [17] and the same year he published his design, Cardoso pointed out that the elements of the Cupid's bow almost always can be found on the medial side of the cleft and simply have to be brought down to a horizontal position [18]. Randall combined the approaches described by Tennison and Cardoso and presented a system of measurements so the vertical height of the Cupid's bow on the normal side could be used to calculate the size of Tennison's triangular flap on the cleft side to achieve symmetry [19].

Millard, working on Korean children when he was on active duty with the U.S. Fleet Marines, conceived and perfected an operation based on an incision locat-

ed where the philtral column should be, carried it across just below the columella, often with a "cutback" incision on the noncleft side [20]. The whole central flap was then rotated down to a normal position and the defect just below the nostril floor was closed with a triangular flap from the lateral cleft margin. This was a great step forward in the cleft lip repair and in Millard's hands, with patient meticulous technique, the results were beautiful. Millard's "rotation advancement" operation was soon the preferred approach for unilateral clefts almost everywhere (Fig. 12.2).

More recently, many steps to "fine tune" these results have been presented. Some objected to carrying the relaxing incision past the midline under the columella and added a very small "z-plasty" just above the vermilion border [21]. This also helped to prevent shortening on the repaired side. Pfeiffer described changing the lip-skin incision to a "wavy line" thus making it less conspicuous. LaRossa curved the incision lines to better reflect the shape of the philtral columns [22, 23]. Mohler noted that most children do not have a shield-shaped philtrum which is the shape produced by the rotation advancement technique, but have philtral columns that are either almost parallel or can actually diverge [24]. He and Salomonson have each described techniques for this situation [25].

It is our feeling that the surgeons doing clefts of the lip should be familiar with a variety of operations and should "fit the operation to the child rather than to fit every child into the same operation" [26].

The skin incisions thus far discussed are mostly concerned with the two dimensions of surface anatomy, and we should not only be concerned with the three dimensional anatomy of the lip but also with the dimension of motion as the lips are highly functional

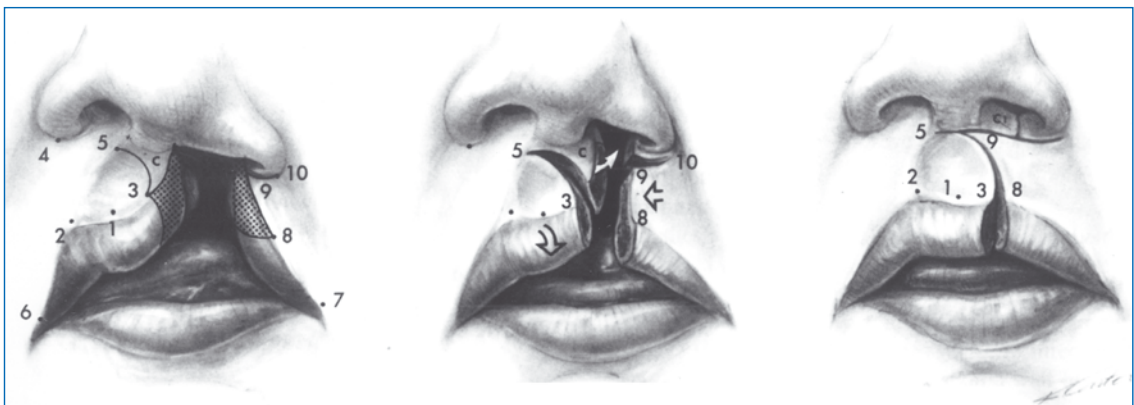


Fig. 12.2. The rotation advancement technique for repair of unilateral clefts. This was first described by D. Ralph Millard, Jr. and was presented at the First International Congress on Plastic Surgery in Stockholm in 1955. It has undergone some

changes since first described, but probably remains the most popular technique for unilateral cleft lip repair the world over. (Illustration with kind permission from [91])

for eating, speaking, and expression, not to mention kissing.

In 1968, Mirislav Fara described the distorted musculature in the lips of stillborn babes with clefts [27]. Briefly, the orientation of this muscle is way out of position. Instead of extending horizontally across the upper lip (with interlacing fibers in the philtrum as described by Latham) in the child with a complete unilateral cleft, the orbicularis muscles fibers roughly parallel the edges of the cleft [28]. They inserted medially near the nasal spine and are laterally directed upward towards the pyriform recess. Randall and Whitaker have pointed out the importance of reorienting the orbicularis muscles if symmetry of shape and function are to be hoped for [29]. Various techniques for repositioning these muscles and reconstructing their anatomy have been presented [30]. GC Park of Seoul, South Korea has pointed out the different functions of the superficial and deep orbicularis muscles and has advocated separate reconstruction of each [31].

In the bilateral cleft, the skin incisions can really be much simpler in that the Cupid's bow is not asymmetrically distorted (unless the clefts are very asymmetrical). The main contention is still whether to save the midline vermillion and have two vermillion incisions extending from the peak of the Cupid's bow on each side or to discard the midline vermillion and reconstruct the vermillion completely with tissue from the lateral elements and a midline vermillion incision.

In complete or near-complete bilateral clefts, there is usually no functioning muscle in the prolabium or midline lip between the philtrum columns. There is a tendency for this midline skin under any circumstances to be stretched and leave a repaired bilateral cleft with a midline philtrum that is far too wide and conspicuously abnormal. For this reason, we try to construct a very narrow philtrum at the initial operation and to suture muscle to muscle across the midline.

In primary nasal tip repair, we mostly follow the teachings of McComb in wide undermining of nasal tip cartilages between cartilage and skin and complete repositioning of the nasal tip cartilages [34]. Matsuo has described intranasal splints or conformers to reshape these nostrils both preoperatively and postoperatively [35, 36]. Mulliken has described reconstructing the nasal tip in bilateral cases, which has made the many steps to reconstruct the short columella unnecessary in most cases. Grayson and Cutting have combined repositioning of alveolar segments with presurgical nasal tip shaping with lovely results [85, 86].

In the 1800s many felt that the cleft in the anterior hard palate and alveolus was not due to a deficiency of tissue, but rather a separation of normal parts. This

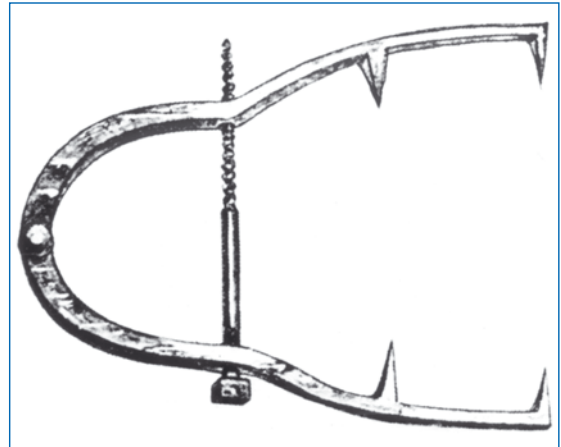


Fig. 12.3. A steel crushing clamp used in infancy by Truman Brophy in 1884 to squeeze the maxillae together, narrowing the cleft in the palate. (From [2], originally from [39])

led to an era of trying to compress these segments together [37]. Some of the techniques, such as described by Roberts, relied on external pressure on the cheeks with compression pieces anchored to a head cap. Others described internal mechanisms, with rather cruel-looking claws impinging on the alveolus and activated by a screw mechanism [38]. Most stressed the desirability to start in the very young infant whose bones were softer and more malleable than the older child (Fig. 12.3).

The approach in the USA was greatly influenced by Dr. Truman Brophy of Chicago. Brophy had a dental degree from the University of Pennsylvania in 1872 and then earned a medical degree from Rush Medical College in Chicago in 1880. The very next year he started the Chicago Dental Institute and became its first Dean – a position he held for the next 39 years [39]. An indication of Brophy's influence in medicine as well as in dentistry was the establishment of the American Association of Plastic Surgeons in Chicago in 1921 as the "American Association of Oral Surgeons." At the insistence of Dr. Robert H. Ivy during its first meeting in Philadelphia that year, the name was changed to the "American Association of Oral and Plastic Surgeons." It was another 20 years before it settled on its present name. The AAPS is regarded as the first society of plastic surgeons in the western hemisphere and probably in the world [40].

Brophy was elected to its presidency not only at this first meeting, but at the next two meetings as well. His influence was widely felt. He started using a system of wire loops and intraoral metal plates for maxillary compression in 1893. (He had probably operated on his first patient using this technique in 1886.) Silver wires were passed through the maxilla and secured

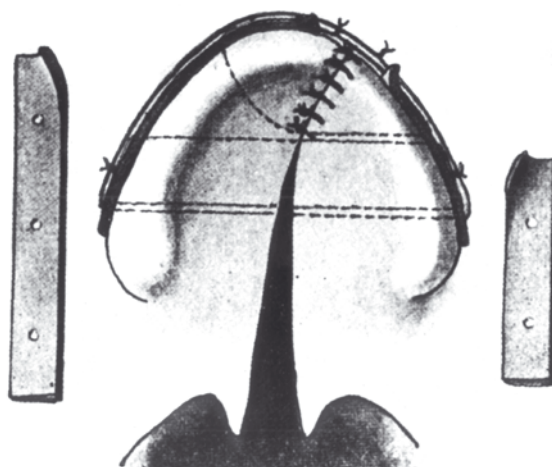


Fig. 12.4. Dr. Truman W. Brophy's ingenious pattern of three silver wire loops passed through the maxillae and threaded through lead plates to selectively reduce the width of the cleft and to align the alveolus. When done "as early after birth as it was possible to operate" while the bones were quite soft, this could be done in one operation. Thomas Graber [42] documents severe facial growth problems in these patients in later life. (From [2], originally in [39])

over lead plates in the buccal sulcus. By a clever design of three such loops, the various parts of the alveolus could be selectively compressed and molded. This was done by selectively twisting the various wires then incising the cleft margins, elevating muco-periosteal flaps, placing horsehair sutures, twisting the wires one more time and tying the sutures. He advocated doing this operation "as early after birth" as is possible to operate" because the neonate's bones are more pliable. Because of his position, publications, and influence, the technique became widely used. In Dr. Brophy's obituary, it is written, "if a path to a desired end were not beaten, he never hesitated to break the virgin soil" [41] (Fig. 12.4).

In 1949 Tom Graber was studying facial growth in children with clefts in the Chicago area [42]. He documented many growth distortions and no doubt was seeing many patients either operated on by Dr. Brophy or one his many disciples.

In 1938, when Dr. Cooper started the Lancaster Cleft Palate Clinic and Dr. Ivy was laying the groundwork for the Pennsylvania State Cleft Palate Program, serious growth disturbances and collapsed alveolar segments were all too common [43]. In 1953, when the senior author started practice, Dr. Wilton Krogman, a physical anthropologist, was getting yearly cephalometric studies on all of our children with clefts starting, if possible, in the first year of life. The senior author was not allowed to operate on a cleft palate until Dr. Krogman told us that the "face was growing well."



Fig. 12.5. A patient with a cleft palate showing bilateral lateral crossbites and crowding, probably due to excessive scar from surgery or some types of crushing maneuver in infancy. Patient of P. Randall

This was usually at 4–5 years of age. The senior author had been trained to close the cleft palate at about 18 months of age. Consequently, our speech results were not very good (Fig. 12.5).

In 1950, Kerr McNeil of Glasgow devised a system of presurgical orthopedic treatment of children with clefts [44]. William Burston soon took up this approach, using treatment "as soon as possible after birth" [45]. A completely new approach to molding the displaced alveolar segments had begun. Burston would obtain the first impression of the upper jaw, preferably within 48 h of birth. A cast would be made from this mold and then would be cut and positioned to passively guide the alveolar segments. Then a bite block was made from this "corrected" plate. The plate was expected to "gag the bite slightly to promote chewing activity." A succession of plates were made (usually four) to achieve the desired positioning. By obturating the hard palate, the plate was expected to help feeding and to keep the tongue out of the cleft. Narrowing of the cleft was also observed. Burston also used external elastic strapping to improve lip position, particularly in the bilateral cleft with a protruding premaxilla.

In 1951, Eduard Schmid reported putting iliac bone grafts into the alveolar area at the time of lip closure, usually at about 8 months of age [46]. This was extended to closure of the anterior hard palate at the same time with further bone grafts to this area.

In 1955, Berg and Johanson with Karl Erik Nordin were doing a preliminary soft tissue approximation of the cleft lip and a mucosal closure of the alveolus and anterior hard palate cleft at 3–4 weeks of age [47]. At this time they also started the "orthopedic correction of the jaw." This was continued until about 6 months of age at which time a careful repair of the cleft lip was done and bone grafts from the tibia were inserted into the alveolar cleft and the anterior hard palate. "Prima-

ry bone grafting" as these procedures came to be known was championed by many including Widmaier, Reichert, Stellmach, Schroeder, Cupar, Pfeifer, Schuchardt, and Nylen to name but a few [48]. In the USA Sheldon Rosenstein, working with Clarence Monroe, Harold Griffith and Bruce Bauer has perhaps the largest series with long-term follow-up cases and recent publications have shown very fine results [49]. In addition, Charles Horton and his group in Norfolk, Virginia, Georgiade and Quinn in Durham, North Carolina, and Brauer and Cronin from Houston, Texas expressed enthusiasm [50].

In 1967, Tord Skoog of Uppsala Sweden reported on raising a large flap of periosteum from the maxilla and moving this to the alveolar cleft and anterior palate. At a second stage, the periosteal flaps are opened and surgical oxidized regenerated cellulose was packed into the cleft, bridging the alveolar gap [51]. This became known as "boneless bone grafting" and indeed bone did form in this area. This was done at the time of lip repair at 3 months of age. Skoog pointed out that regenerated bone from transplanted periosteum "grows with the individual, unlike the static transplanted bone." Joss in Norwich, England, O'Brien in Melbourne, Australia, and Rintala in Helsinki, Finland became enthusiastic followers [52]. In some cases the Surgicel (Johnson & Johnson) was inserted at a second stage, in some at the first stage, and in some it was not used at all – with little difference in the results. Rintala reported on 63 patients in 1974. Good bone was found in 54% of the cases, "diffuse" bone in 22%, and no bone in 24% – "regardless of the Surgicel" [53].

It was felt that perhaps Skoog's periosteal flap had such a poor blood supply that the transfer would succeed even as a free flap. In 1969, Millard in questioning this thought designed an experiment in rabbits, comparing burr holes in cranium, half with a free periosteal graft, covered by a periosteal flap and half with only the dura in continuity. The latter did form some bone in two of nine rabbits, but it was very thin. In the grafted sites (in those that either did not become infected or did not die) considerably more bone was produced [54].

In fact, Veiju Ritsila of Helsinki, Finland working with Arne Rintala showed that in patients with free tibial periosteal grafts compared with those done with the Skoog maxillary periosteal flap, the free tibial periosteum produced better bone. In 63 patients treated using Skoog's technique, 54% had a definite bone bridge, 22% had diffuse ossification, and in 24% there was no bone formation. In the 25 free tibial periosteal grafts, the figures were 76%, 12%, and 12% [55].

With these approaches providing bone in the alveolar cleft and often the anterior hard palate as well, enthusiasm continued to increase. Yet Sam Pruzansky

in the USA remained unconvinced and was often very critical [88]. In a very large series without primary bone grafts he found "37% with no crossbite, 40% with complete crossbite on the affected side and the rest with varying degrees of incomplete crossbites. Since a significant number of patients do not develop crossbite at all, is there justification for treating all patients by presurgical maxillary orthopedics and bone grafting? In our judgment there is none. Moreover, the malocclusion present in the preschool child can be readily, quickly and less expensively treated by simple expansion procedures."

Alfred Rehrman in 1969 presented his findings on the longest study between surgery and re-examination [57]. Having worked with Schuchardt; he did his studies in Düsseldorf with non-bone grafted controls and concluded that bone grafting did not bring about permanent stabilization of the segments. He said, "Early bone grafting in nearly all of our cases provoked retardation in development of the maxillary arch and local arrest of maxillary bone. For this reason, we have abandoned primary and early secondary bone grafting and limited osteoplasty to the time after the secondary dentition."

Huddart in England compared children who had had presurgical orthopedics with those who had not and in 1966 "found no apparent difference in the two groups" [58]. Dennis Glass in England in 1970 had similar findings [89]. Bill Olin in the USA concluded in 1978 "the procedure was not indicated in most cases" [59].

In 1965, Johansen in reviewing 100 of his earliest cases concluded that "the results were so disappointing that (he) has discontinued this method of treatment. [90]. Only at 12–18 years of age did he bone graft the alveolus. In 1969 he said "I know in documenting the series which now parallels the bone grafted series that our results are much better today, without the primary bone graft, than they were when we started." [60]. Others agreed including Wilfried Schilli in Freiburg and Rudi Stellmach of Berlin [54, 55].

In other studies on Skoog's "boneless bone grafting," a sizeable series from Andersen's clinic in Copenhagen by Peter Holm concluded, "Surgical procedures that result in bone formation across the cleft should be abandoned." [61, 62]

In 1970, Ralph Latham was working at Duke University with Nicholas Georgiade. They developed [63, 64]. The developed a palatally pinned apparatus to expand the maxillary segments and to retrude the protruding premaxilla at birth [63, 64]. Millard was cautious about relying on the constricting force of a repaired cleft lip to mold the bony elements. "Tension on a surgical repair usually led to thickened scar tissue." He said, "I particularly resent having my lip scar shoulder any unnecessary stress or strain." [65]

Latham, whose home base was the University of Western Ontario, felt that he could be very helpful in aligning these structures, while Millard's interest in closing the alveolar cleft using gingival mucosa and periosteum on the oral side and tissue from the border of the lip cleft on the nasal side would be greatly benefited by minimizing the alveolar gap [65]. It was Latham's aim to bring the alveolar segments within 2 mm of each other preoperatively. Further, if this could be done before lip closure, access to this difficult area would be greatly improved. In 1976 they joined forces and did their first gingival periosteoplasty on a 5-month-old infant with a complete unilateral cleft, closing the alveolar cleft, closing the nasal floor and carrying out a lip adhesion operation [65]. Postoperatively the dental arch "must be expanded and retained with appliances until the cleft has filled with new bone that can maintain the dimensions of the palate." Millard has continued to be very enthusiastic about this combined approach of Latham's preoperative positioning, alveolar periosteoplasty, and postoperative retention.

Finally, after 20 years of collecting data Berkowitz in 2004 published a 40-year comparative study of the Latham-Millard treatment with a conservative non-orthopedic and non-periosteoplasty using delayed secondary alveolar bone grafting in complete unilateral and bilateral clefts of the lip and palate. The results unequivocally showed that presurgical orthopedic manipulation with periosteoplasty caused severe midfacial retrusion with anterior crossbites which are difficult to correct (see Chapter 20) [66]. In CUCPL and CBCLP cases the retracted premaxilla became fused with the posterior segments, closing off the cleft spaces creating a recessive midface. The bony bridge prevents the easy correction of the buccal and/or anterior crossbites. Orthodontic correction often extends into adolescence and in some instances necessitates additional surgery.

Meanwhile other clinics are also using presurgical orthopedics and periosteoplasty, however, even though it is being in use for a numbers of years no outcome studies have been reported [85].

While presurgical orthopedics and "primary bone grafts" were declining in popularity, "secondary bone grafting" at 9–11 years of age was gaining attention. Abyholm and Bergland in Oslo were showing many of their cases operated on in their mixed dentition with spectacular results, i.e., beautiful gingival reconstruction achieved with gingival flaps and bone grafts and remarkably good occlusions with permanent teeth erupting through the bone grafts in good occlusion [67]. LaRossa, Buchman, et al. have shown that this can be achieved with iliac bone, cranial bone, or rib, but that success can be expected more with iliac bone and least with rib [68].

In 1965, Sheldon Rosenstein was part of the team at Children's Memorial Hospital in Chicago. Working at first with Clarence Monroe then Desmond Kernahan, Harold Griffith, and Bruce Bauer, they have a rather remarkable series of cases, well studied with as much as a 35-year follow-up of 135 patients who had early bone grafting [69].

A maxillary prosthesis is placed before lip closure. Lip closure is done at 6–8 weeks of age. Molding is then carried out under the repaired lip until the alveolar segments are in "butt alignment." At this time, usually 4–8 months of age, a *small* autogenous rib graft is placed across the cleft maxillary alveolus in a subperiosteal pocket. The appliance is continued for 6–8 weeks, after which it might be used only to assist in feeding. The rest of the palatal cleft is then closed before 1 year of age with an intravelar veloplasty. His results are outstanding.

Eppley, in discussing Rosenstein's 2003 paper, noted that it had been shown that "extensive early bone grafting across and around the premaxillary suture does result in long-term maxillary growth inhibition" [70]. Rosenstein has carefully studied his results and compared them with RE Long of the Lancaster Cleft Palate Clinic and Bruce Ross at the Toronto Hospital for Sick Children and has documented very favorable outcomes in maxillary growth, tooth preservation and stabilization, occlusion, the need for secondary surgery, the incidence of fistulae, and speech outcome [70, 71].

Barry Eppley, working with Michael Sadove, reported similar findings in 350 cases [72]. He stresses the need for good end-to-end alignment and a small graft with minimal undermining. "If good alignment between the lesser and greater segments cannot be obtained or the residual width between the segments is two millimeters or greater, bone graft incorporation drops precipitously."

Grayson and Cutting at New York University have an approach which seems to be based on the same principles. They have not as yet published any occlusal and facial outcome studies [85, 86].

A key part of our past is the history of the record keepers, those who create and maintain longitudinal studies. Only by looking at our long-term results can we evaluate our treatment plans. Sam Pruzansky was one of our first and most ardent record keepers. He would continually berate those of us who did not keep appropriate records, particularly if we drew conclusions concerning choices of treatment on scant evidence. Prunzansky and Aduss published many longitudinal studies on all aspects of cleft palate treatment starting at birth and extending through adolescence. Sam Berkowitz, one of Prunzansky's students, continued this tradition by emphasizing the need to back up all surgical concepts with objective records. Unfor-

tunately this is not being done in U.S. except at the University of Illinois Craniofacial Center. Berkowitz strongly urged that since bone is missing in the cleft spaces the space needs to be left open by keeping the alveolar segments out of contact this upper to lower anterior arch congruency would result without preventing anterior crossbites developing. He strongly cautioned against using early bone grafting or periosteoplasty.

The Lancaster Cleft Palate Clinic started the team approach to cleft care under Dr. Herbert Cooper in 1938. Backed by the local Rotary Club, Cooper literally housed his patients in his own home while getting them into speech therapy, beginning their orthodontic and prosthodontic treatment, and scheduling them for appropriate surgery. After retirement from Children's Hospital of Philadelphia, Wilton Krogman joined the Lancaster Cleft Palate team and as a physical anthropologist augmented their longitudinal studies [74]. The team at Iowa City and a number of other teams did similar work [75].

In 1938, Robert H. Ivy started working with the State Department of Health in Pennsylvania [76]. He eventually put together about thirteen multidisciplinary teams for the team care of children with clefts in that state. The "team approach" has now become the accepted pattern of care for children with clefts throughout the industrialized world. In fact, it is now the preferred approach for many other problems such as breast malignancy, myelomeningocele, juvenile diabetes, Down's syndrome, 22q11 syndrome, etc.

Leslie Farkas at the Toronto Hospital for Sick Children was particularly interested in studying and recording the faces of children with clefts [77]. Bruce Rose in the same institution collected data from many places on facial bone growth and palatal architecture [78]. Our own editor and author, Sam Berkowitz, not only added novel ways of recording palatal anatomy and contour, but also is now continuing one of the best series on long-term results [79].

In addition, there are a number of clinics where several generations of specialists have carried on excellent traditions. This list is very incomplete but one is reminded of two generations of Fogh-Andersen in Copenhagen; two generations of Schweckendieks in Marburg; two generations of Skoogs in Uppsala; and the legacies of Blair, Brown, McDowell, and Marsh in St. Louis, Missouri; Veau, Petit, and Malek in Paris; and Priestersbach, Morris, and Karnell in Iowa City, Iowa. This list could go on and on. Suffice it to say, we owe a great debt to those who have gone before us.

We are not knowledgeable enough to evaluate the many fine cleft palate clinics in Europe and Asia. Surely the studies by the team at the Karolinska Institute in Stockholm; the team started by Arni Rintala in Helsinki; the work of Margaret Hotz, her husband

Rudolph and Wanda Gnoinski of Zurich, Switzerland; the studies of Rehrman in Düsseldorf, Germany; and the studies of adults with unoperated clefts by Ortiz-Monastario of Mexico City, Hans Friede, Jan Lilja and Bengt Johansten of Gotemborg-Sweden; Gunvor Semb and Bergland (deceased) of Oslo-Norwy; Samir Bisahra, Bill Olin, J. Bardach (deceased) from Illinois, USA; J.D. Subtelny and J.A. Subtelny from Rochester, USA; are but a few of the many researchers contributing to this field.

McNeil in England started people thinking about early orthopedic manipulation of the alveolar segments. In 1989 at the International Congress in Jerusalem, Bill Shaw pointed out that "very few papers on cleft palate repair were designed as scientific studies with suitable controls and statistical analyses showing significant results" [80]. He further stated that "if good multi-institutional studies can be designed to research the treatment of conditions as variable as cancer, we should be able to do the same for cleft palate" [80]. Shaw has done just this in organizing Eurocleft studies for the purpose of critically evaluating results, documenting case histories and structuring collaborative studies between institutions [81]. The work by Sam Noordhoff in Taiwan, Matsuo in Japan, Grayson and Cutting in New York, Genecov and Salyer in Dallas as well as many others has further developed this field [82-84]. One of the exceptional contributions to the field of clefts is the unusual three-volume set of books called, "Cleft Craft," by Dr. D. Ralph Millard [84]. For completeness of history, developments, contributions, and humor combined with his exceptional insight, there is no peer, and without these volumes, it would have been difficult to write this brief review. Answers are a long time in coming, but with cooperation, documentation, and constructive criticism, we seem to be making progress.

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Core Curriculum for Cleft Lip/Palate and other Craniofacial Anomalies

A Guide for Educators
prepared by the



American Cleft Palate –
Craniofacial Association

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CONTRIBUTORS

Isaac L. Wornom, III, MD, Editor
Leslie A. Will, DMD, MSD, Co-Editor

Alphonse R. Burdi, PhD, Anatomy
Samuel Berkowitz, DDS, MS, Orthodontics
Mary L. Breen, MS, RN, Nursing
Noreen Clarke-Sheehan, MSN, RN, Nursing
Virginia M. Curtin, RN, MS, PNP, Nursing
Linda L. D'Antonio, PhD,
Speech-Language Pathology
Craig D. Friedman, MD,
Otolaryngology/Head & Neck Surgery
Ann Tucker Gleason, PhD, CCC-A, Audiology
Donald V. Huebener, DDS, MS, Pediatric Dentistry
Marilyn C. Jones, MD, Pediatrics
Austin I. Mehrhof, DDS, MD, Plastic Surgery
Sharron A. Newton, BSN, Nursing
Lynn C. Richman, PhD, Psychology
John E. Riski, PhD, Speech-Language Pathology
R. Bruce Ross, DDS, MSc, Orthodontics
James D. Sidman, MD,

Otolaryngology/Head & Neck Surgery
Barry Steinberg, PhD, DDS, MD,
Oral/Maxillofacial Surgery
Sandra Sulprizio, MSPA, Speech-Language Pathology
Ruth Trivelpiece, MEd, Speech-Language Pathology
Kim S. Uhrich, MSW, CCSW, Social Work
Carol R. Ursich, BSN, Nursing
Linda D. Vallino-Napoli, PhD, Speech-Language
Pathology
Leslie A. Will, DMD, MSD, Orthodontics
Isaac L. Wornom, III, MD, Plastic Surgery

EDITED BY THE 2002–2003 EDUCATION COMMITTEE

Isaac L. Wornom, III, MD, Chair
Samuel Berkowitz, DDS, MS
Alphonse R. Burdi, PhD
Charles L. Castiglione, MD
Adriana C. Da Silveira, DDS, PhD
Jessica DerMarderosian, BA
Amelia F. Drake, MD
Robin A. Dyleski, MD
Susan E. Eastwood, MSc
Jaime Gateno, DDS, MD
Mary J. Hauk, DDS
William Y. Hoffman, MD
Bruce B. Horswell, DDS, MD
Katherine A. Kelly, DDS, PhD
L. Elizabeth Peterson, MD
Joan T. Richtsmeier, PhD
Iris H. Sageser, RDH, MS
Sandra Sulprizio, MSPA
Ruth Trivelpiece, MEd
Leslie A. Will, DMD, MSD
Jack C. Yu, DMD, MD

Virginia A. Hinton, PhD, Council Liaison

The ACPA Core Curriculum will be updated and enhanced on a regular basis. Readers are encouraged to find the most recent version of this document on the ACPA website at www.acpa-cpf.org.
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TABLE OF CONTENTS

**Core Curriculum for Cleft Lip/Palate
and other Craniofacial Anomalies**

CONTRIBUTORS	283
--------------------	-----

Core Curriculum Outline	285
-------------------------------	-----

Section 1**Interdisciplinary Team Care, Classification,**

Airway, and Feeding	285
----------------------------------	------------

I. Team Evaluation	285
--------------------------	-----

Airway Maintenance	285
--------------------------	-----

Feeding and Nutrition	286
-----------------------------	-----

II. Classification and Anatomy	285
--------------------------------------	-----

III. Airway and Feeding	285
-------------------------------	-----

Airway Maintenance	286
--------------------------	-----

Feeding and Nutrition	286
-----------------------------	-----

IV. Noncleft Craniofacial Anomalies	286
---	-----

Hemifacial Microsomia	286
-----------------------------	-----

Craniosynostosis	287
------------------------	-----

Other Craniofacial Anomalies	287
------------------------------------	-----

V. Craniofacial Development	287
-----------------------------------	-----

Developmental Craniofacial Biology	287
--	-----

Craniofacial Embryology	287
-------------------------------	-----

Basic definitions in craniofacial biology ...	288
---	-----

SECTION II**General Role of the Various Disciplines****in Treating Patients with Clefts**

and Craniofacial Anomalies	289
---	------------

I. Audiology	289
--------------------	-----

Early Identification	289
----------------------------	-----

Management	289
------------------	-----

Monitoring	289
------------------	-----

II. Genetics	290
--------------------	-----

Cleft lip with or without cleft palate	290
--	-----

Cleft palate alone	291
--------------------------	-----

Non-cleft craniofacial abnormalities	291
--	-----

III. Nursing	291
--------------------	-----

Prenatal	291
----------------	-----

Neonatal	291
----------------	-----

Infant/Toddler	291
----------------------	-----

Preschool/School-Aged/Adolescents	291
---	-----

IV. Oral and Maxillofacial Surgery	292
--	-----

Prenatal	292
----------------	-----

Neonatal	292
----------------	-----

Infant	292
--------------	-----

Toddler	292
---------------	-----

Preschool	292
-----------------	-----

School-Aged	292
-------------------	-----

Adolescents	292
-------------------	-----

V. Orthodontics	292
-----------------------	-----

Prenatal	292
----------------	-----

Neonatal	292
----------------	-----

Infant	292
--------------	-----

Toddler	292
---------------	-----

Preschool	292
-----------------	-----

School-Aged	293
-------------------	-----

Adolescents	293
-------------------	-----

Adult	293
-------------	-----

Record keeping	293
----------------------	-----

VI. Otolaryngology - Head and Neck Surgery ..	293
---	-----

Prenatal	293
----------------	-----

Neonatal	294
----------------	-----

Toddler	294
---------------	-----

Preschool	294
-----------------	-----

School-Aged	294
-------------------	-----

Adolescent and Adult	294
----------------------------	-----

VII. Pediatric Dentistry	294
--------------------------------	-----

Prenatal	294
----------------	-----

Neonatal	294
----------------	-----

Infant	294
--------------	-----

Toddler	294
---------------	-----

Preschool	294
-----------------	-----

School-Aged	294
-------------------	-----

Adolescents	294
-------------------	-----

Adult	295
-------------	-----

VIII. Plastic Surgery	295
-----------------------------	-----

Prenatal	295
----------------	-----

Neonatal	295
----------------	-----

Infant	295
--------------	-----

Toddler	295
---------------	-----

Preschool	295
-----------------	-----

School-Aged	295
-------------------	-----

Adolescents	296
-------------------	-----

Adult	296
-------------	-----

IX. Psychology and Clinical Social Work	296
---	-----

Prenatal	296
----------------	-----

Neonatal	296
----------------	-----

Infant	296
--------------	-----

Toddler	296
---------------	-----

Preschool Development	296
-----------------------------	-----

School-Aged Child	296
-------------------------	-----

Adolescents	297
-------------------	-----

Adulthood	297
-----------------	-----

X. Speech and Language Pathology	297
--	-----

Neonatal and Infancy	297
----------------------------	-----

Toddler	297
---------------	-----

Preschool/School-Aged/Adult	298
-----------------------------------	-----

References	299
------------------	-----

Core Curriculum Outline

The American Cleft Palate-Craniofacial Association (ACPA) believes that children born with clefts and other craniofacial anomalies are provided optimum care when they are assessed and treated by a team of specialists with expertise in a variety of areas. Health care specialties involved with the care of clefts and other craniofacial anomalies include audiology, genetics, nursing, oral and maxillofacial surgery, orthodontics, otolaryngology/head and neck surgery, pediatric dentistry, plastic surgery, psychology and clinical social work, and speech-language pathology.

This core curriculum was created by the Education Committee of ACPA to be used as a guide for educators in these various disciplines, when planning the essential parts of their curriculum related to cleft and craniofacial anomalies. It was developed after a survey by ACPA of educators in these disciplines showed a need for such an outline. The Core Curriculum is not intended to cover all possible aspects of cleft and craniofacial management. Rather, it is intended to provide an outline of services that are appropriate for most children affected by these disorders.

The Core Curriculum is divided into two broad sections. The first covers the basics of interdisciplinary team care, classification of craniofacial anomalies, craniofacial development and etiology. The second section covers the role of each discipline in the care of a patient with a cleft or craniofacial anomaly. It is organized by patient age, within each discipline, and covers the essential aspects and knowledge bases that are essential for providing adequate care. Just as in a team there may be overlap between specialists in their observations, core knowledge and treatment expertise, this core curriculum reflects some overlap between specialties in these areas.

Section 1 Interdisciplinary Team Care, Classification, Airway, and Feeding

I. Team Evaluation

The initial evaluation of the patient should be by a pediatrician, who is knowledgeable about all aspects of the infant's care. Optimum management of children with clefts and craniofacial anomalies is provided by a team of health care professionals with a specific interest in these anomalies. Team evaluation should be performed early in life and, ideally, the initial contact with the team should be prior to the infant's discharge from the hospital following birth. This allows the parents to receive information about their baby's

problem and subsequent treatment, as soon as possible. Team members include specialists from:

- A. Audiology
- B. Genetics
- C. Nursing
- D. Oral and maxillofacial surgery
- E. Orthodontics
- F. Otolaryngology and head and neck surgery
- G. Pediatric dentistry
- H. Plastic surgery
- I. Psychology and clinical social work
- J. Speech-language pathology

II. Classification and Anatomy

The cleft or craniofacial anomaly is usually classified during the initial examination of the infant. Craniofacial anomalies, other than clefts, are discussed in Section IV.

- A. Clefts of the lip and clefts of the palate can occur simultaneously or separately.
- B. The most common classification system for clefting uses the terms primary and secondary palate to define the cleft.
- C. The dividing point of the primary and secondary palate is the incisive foramen. The primary palate is anterior to this anatomic point and the secondary palate is posterior to it.
- D. The primary palate includes:
 - 1. Lip
 - 2. Alveolus
- E. The secondary palate includes:
 - 1. Hard palate
 - 2. Soft palate
 - 3. Uvula
- F. Any cleft of the primary or secondary palate may be complete or incomplete, depending on whether or not the cleft involves the entire anatomic structure.
- G. Any cleft of the primary or secondary palate may be unilateral or bilateral.
- H. Submucous clefts of the secondary palate may also occur. These can be detected by visual inspection, ultrasonography or radiography.

III. Airway and Feeding

As with any newborn, the primary concerns in the neonatal period are airway maintenance, breathing, and feeding. Some of the anatomic variations in children with clefts and craniofacial anomalies may have an impact on these functions.

A. Airway maintenance

1. Cleft lip and/or cleft palate rarely cause problems with the upper airway or breathing, when there are no other associated problems.
2. Pierre-Robin Sequence is the most common anatomic deviation associated with clefting that can result in airway and breathing problems.
 - a. Pierre-Robin Sequence results in a combination of malformations, consisting of mandibular hypoplasia, glossoptosis, and midline cleft of the secondary palate.
 - b. Usually airway problems can be managed with prone positioning and time for growth to occur. However, dental prosthetic or surgical intervention, including tracheostomy, may be required in severe cases. In addition, distraction osteogenesis of the mandible has been used to treat some infants with severe airway problems due to Pierre Robin Sequence, but its use at this age is controversial.
3. Craniofacial anomalies can also be associated with airway problems. These include, but are not limited, to:
 - a. Syndromal craniosynostosis with severe mid-face hypoplasia.
 - b. Any syndrome associated with a severely deficient mandible, such as severe Treacher-Collins Syndrome.
 - c. Choanal atresia.

B. Feeding and Nutrition

1. Babies with isolated clefts of the lip and/or palate can usually feed by mouth with some adjustments to bottle-feeding techniques. Tube feeding is rarely required.
2. Babies with isolated cleft lip may be able to breast feed, but it is unlikely that a child with a cleft palate will be able to successfully breast feed because of nasal spillage and the difficulty in maintaining an adequate suction.
3. Despite the problems with maintaining sucking pressures, the swallowing mechanisms in children with cleft palate are usually normal. Therefore, if the milk or formula can reach the oropharynx, the natural swallowing reflexes can move it into the esophagus.
4. Some nasal regurgitation may occur, but this is rarely more than an inconvenience. Upright positioning during feeding may help reduce the occurrence of nasal regurgitation.
5. The strategies that have been developed to feed infants with clefts of the palate are designed to overcome the lack of negative pressure developed during sucking. These include, but are not limited, to:
 - a. Cross-cutting fissured nipples.

- b. Squeezing a soft bottle to help with the flow of milk.
- c. Pumping the breasts to deliver breast milk via bottle.
- d. Developing patience in feeding.
- e. Feeding instruction and follow-up with a feeding specialist on the cleft palate team.
6. It is important to ensure that the energy that a child expends during feeding does not exceed the nutritional and caloric intake from the feeding. This problem may occur if feeding takes more than 30 minutes.
7. Steady weight gain is the most important indicator of adequate food intake. Close follow-up with a pediatrician or other health care provider is necessary to ensure that consistent weight gain is achieved.
8. Frequently, airway problems will be exacerbated during feeding. The combination of the inability to maintain adequate sucking and airway problems may lead to the need for an alternative feeding method.
9. These same principles apply to babies with other craniofacial anomalies, even though the anatomic cause of their feeding problems may be mandibular or maxillary hypoplasia, rather than clefting.

IV. Noncleft Craniofacial Anomalies

A. Hemifacial Microsomia

1. This is the second most common congenital anomaly of the head and neck after clefting. Hemifacial microsomia includes:
 - a. Malformation of the external ear with varying degrees of microtia and/or other ear anomalies.
 - b. Malformation of the mandible, with varying degrees of shortening or absence of the ramus of the mandible. Subsequent chin deviation and malocclusion will occur.
 - c. Varying degrees of maxillary hypoplasia.
 - d. Facial nerve weakness or absence in severe cases.
2. Team management typically includes:
 - a. Protection of hearing in the normal ear.
 - b. Orthodontic management, combined with rib graft reconstruction or distraction osteogenesis of the ramus, at age 4 to 6 years.
 - c. Ear reconstruction with otoplasty or rib cartilage, depending on the severity of the anomaly.
 - d. Orthognathic surgery and additional orthodontic management after facial growth is complete.

B. Craniosynostosis

1. Craniosynostosis is early fusion of the sutures between the bones of the skull where growth naturally occurs, thus, precluding growth at the suture site.
2. It can occur in isolation or as a part of several syndromes.
3. Non-syndromal craniosynostosis is classified morphologically by the suture involved and subsequent skull shape.
 - a. One or two sutures involved with different skull and upper face deformities depending on suture.
 - 1) Sagittal suture-scapocephaly
 - 2) Unicoronal suture-plagiocephaly
 - 3) Metopic suture-trigonocephaly
 - 4) Bicoronal sutures-brachycephaly or turriccephaly or both
 - b. Increased intracranial pressure and developmental delay is rare.
 - c. Correction usually requires one operation in infancy. Secondary surgery is uncommon.
4. Syndromal craniosynostosis is classified according to the name of the syndrome.
 - a. Turribrachycephalic skull shape is common.
 - b. Five syndromes of which craniosynostosis is a part
 - 1) Crouzon syndrome
 - 2) Apert syndrome
 - 3) Carpenter syndrome
 - 4) Saethre-Chotzen syndrome
 - 5) Pfeiffer syndrome
 - c. All are inherited in an autosomal dominant fashion, except Carpenter syndrome, which is recessive.
 - d. Syndactyly of the hands and feet is part of Apert, Carpenter, and Pfeiffer syndromes.
 - e. Increased intracranial pressure and developmental delay are more common than in non-syndromal craniosynostosis, but are not universal.
 - f. Multiple operations throughout life are usually required to treat these patients.

C. Other Craniofacial Anomalies

1. Orbital hypertelorism
 - a. Orbits laterally displaced making the eyes appear too far apart.
 - b. Caused by nasofrontal dysplasia, encephalocele, tumor, or complex craniofacial clefts.
 - c. Can be corrected with surgery.
2. Treacher-Collins Syndrome
 - a. Autosomal dominant.
 - b. Includes varying degrees of zygomatic hypoplasia, lower eyelid coloboma, mandibular hypoplasia, and microtia.

- c. Multiple surgeries throughout life required to treat.

3. Craniofacial clefts

- a. Clefting can occur in the upper face and forehead, as it does in the lip and palate, and can involve all anatomic layers including bone.
- b. These clefts are very rare, may be very deforming, and may require multiple surgeries to treat.

V. Craniofacial Development

A. Developmental Craniofacial Biology

1. Molecular regulation of craniofacial morphogenesis
 - a. Facial rhombomeres, HOX and OTX2 genes
 - b. Patterns of neural crest formation, migration, fates.
 - c. Abnormal neural crest development (neuro-cristopathies), e.g., Treacher Collins syndrome (mandibulofacial dysostosis), Pierre Robin sequence, DiGeorge sequence, Hemifacial Microsomia.
 - d. Molecular regulation of skeletal morphogenesis, e.g., Fibroblast growth factors (FGFs) and receptors (FGFRs).
 - 1) Fibroblast Growth Factor (FGF) gene
 - 2) Fibroblast Growth Factor Receptor (FGFR) gene
 - e. Molecular regulation of eye development, e.g., PAX6, PAX2, BMP7 genes, and sonic hedgehog (shh).
 - 1) PAX6 gene
 - 2) PAX2 gene
 - 3) Sonic hedgehog (shh)
 - f. Molecular regulation of palate formation, e.g. Epidermal growth factor (EGF), transforming growth factor- α (TGF α).
 - g. Genes and tissue interactions in tooth development.
 - h. "Time table"
 - 1) Chronology in craniofacial embryology
 - 2) Critical periods of peak morphogenesis

B. Craniofacial Embryology

1. Development of skull
 - a. Neurocranium: membranous (desmocranium) and cartilaginous (chondrocranium).
 - b. Viscerocranium, e.g. maxilla, palate, mandible.
 - c. Morphogenesis of sutures and synchondroses.
2. Development of the face, eye, nose, lip, palate, tongue, ear
 - a. Facial prominences (5).
 - b. Roles of olfactory, optic and otic placodes.

- c. Primary and secondary palates.
- d. Pharyngeal arch contributions to tongue formation.
- e. Morphogenesis of the ear – internal, middle, and external ears.
- f. Morphogenesis of the eye
 - 1) Optic cup and lens
 - 2) Retina, lens, iris and ciliary body
 - 3) Choroid, sclera and cornea
 - 4) Optic nerve
- g. Organization and development of orofacial and tongue muscles.
- h. Morphogenesis of velopharyngeal muscles.
3. Pharyngeal arches and pouches
 - a. Organization of arches and pouches.
 - b. Component tissues of arches and pouches.
 - c. Structures developing from arches and pouches.
 - d. Malformations related to abnormal pharyngeal arch and pouch formation, e.g., branchial fistulae and cysts.
4. Development of teeth
 - a. Stages in typical tooth morphogenesis.
 - b. Tissue interactions in tooth development.
 - c. Development and plan of primary, mixed, and permanent teeth.
- C. Basic definitions in craniofacial biology
 1. Anomaly (Major): Condition often defined as malformations (or defects) that create significant medical problems and require surgical and medical management.
 2. Anomaly (Minor): Condition often described as morphologic features that vary from those that are most commonly seen in the normal population but, in and of themselves, are not associated with increased morbidity.
 3. Association: A group of anomalies that occur more frequently together than would be expected by chance alone but do not have a predictable pattern or unified etiology.
 4. Critical Periods: Intrauterine (chiefly embryonic) periods of peak organogenesis during which time the embryo is at high risk for teratogen exposure.
 5. Disruptions: A condition where a fetal structure is growing normally and then growth is arrested by a factor(s) that disrupts the normal development process.
 6. Deformation: A condition (often temporary) caused by an abnormal external force on the fetus during in utero development that results in abnormal form and growth of the fetal structure or region.
 7. Dysplasia: Anomalous development related to an underlying tissue disturbance where the cellular architecture or growth of a tissue is not normally maintained throughout development.
 8. Ectoderm: Outermost of the 3 primary layers that forms the nervous system and outer skin (epidermis).
 9. Endoderm: Innermost of the 3 primary layers that forms the lining of the gut.
 10. Etiology: Underlying factors and causes for congenital anomalies or birth defects. Note that the same apparent conditions may have different etiologies in different individuals.
 11. Facial Prominences: These are the five major building-block structures (frontonasal [1], maxillary [2], and mandibular [3]) that play important roles in the formation of the embryonic head and face.
 12. Fibroblast Growth Factor (FGF): A family of signaling key roles in embryogenesis, including that of the limbs, skeleton, and head and face.
 13. Fibroblast Growth Factor Receptor (FGFR): Protein receptor sites located on cell membranes which bind with specific signaling molecules (e.g. Shh, FGF) that transmit molecular signals to the cell nucleus and the specific development of the cell(s).
 14. Field Defects: Term often used to describe related malformations in a particular region and sometimes used interchangeably with the term “sequence”.
 15. Genotype: This is the fundamental genetic constitution or composition of an individual.
 16. HOX genes: A set of homeobox genes with identified DNA sequences controlling those that play important roles in morphogenesis of the body, in general, and in specific structures of the head and face.
 17. Malformation: Malformation signifies that fetal development and growth did not progress normally due to underlying genetic, epigenetic, or environmental factors that altered development of a specific structure or structures.
 18. Mesoderm: Middle layer of the 3 primary layers that forms the dermis, bone, cartilage, blood vessels and connective tissue.
 19. Neural Crest: Layer of cells superior to the developing neural tube that migrate to become part of nearly all major structures and organ systems in the body.
 20. OTX gene: A homeobox containing gene that plays an important role along with HOX genes in the embryogenesis of the brain and the morphogenesis of the first pharyngeal arch and its derivatives, especially the craniofacial regions.
 21. Pathogenesis: The cellular basis of abnormal development associated with known or hypothesized etiologies.

22. **Pharyngeal Arches:** Paired arches in embryonic neck region separated by pharyngeal grooves which play important roles in development of the head and neck.
23. **Pharyngeal Grooves:** Deep depressions between pharyngeal arches in embryonic neck region. The first groove persists and forms the external acoustic meatus.
24. **Pharyngeal Pouches:** Outpocketings from the embryonic pharynx wall that play important roles in development of structures, such as the tympanic membrane, tonsils, thymus and parathyroid glands.
25. **Phenotype:** Phenotype is the observed result of the interaction of the genotype with environmental factors, i.e. the observable expressions of a particular gene or genes.
26. **PAX gene:** The PAX gene family (e.g. PAX2, PAX6) is an important group of genes that play key roles in the morphogenesis of such structures as the ear, eye, and nose.
27. **Rhombomeres:** Blocks of tissue located lateral to the embryonic hindbrain (rhombencephalon), which provide for fundamental organization of hindbrain and eventually play key roles in facial development.
28. **Sequence:** A group of related anomalies that generally stem from a single initial major anomaly that alters the development of other surrounding or related tissues and structures.
29. **Sonic Hedgehog (shh):** A protein “signaling” molecule that plays the most important role in shaping the entire embryo, and in specific structures of the head and face, including teeth.
30. **Syndrome:** A condition generally recognized and defined as a well characterized constellation of major and minor anomalies that occur together in a predictable fashion presumably due to a single underlying etiology (e.g. genes, chromosomes, teratogens).

SECTION II

General Role of the Various Disciplines in Treating Patients with Clefts and Craniofacial Anomalies

I. Audiology

The audiologist on the cleft and craniofacial team provides information regarding hearing sensitivity and mechanical function of the ears. Many syndromes with cleft lip and palate as a feature also have a risk for hearing loss. In addition, the function of the Eustachian tube (which connects the space behind the eardrum to the back of the throat) may be impaired by

the cleft of the palate, putting the patient at increased risk for frequent ear infections. Stable hearing sensitivity is required for the proper development of speech and language. Children with cleft lip and palate are already at risk for speech and language problems due to anatomic abnormalities of the “articulators.” For this reason, it is important to identify hearing loss early by monitoring their auditory sensitivity on a regular basis, in order to minimize complications of abnormal or fluctuating hearing on speech development.

A. Early Identification

1. **Newborn hearing screening:** Techniques have been developed to test hearing regardless of age. Hospitals in many states routinely screen the hearing of all newborns. Diagnostic testing is performed in cases where the infant does not pass the screening test.
2. **High-risk testing:** In states where universal screening is not available, children with craniofacial anomalies are tested because of their high risk for hearing loss status. Although specific test protocols will vary from one facility to another, “high risk” infants should be tested prior to age 4 months.
3. **Diagnostic testing:** In cases where the infant does not pass the screening test, diagnostic testing will be performed in order to determine the severity of hearing loss as well as the type of hearing loss (“nerve deafness” vs. hearing loss due to ear infection), and whether the hearing loss is in one ear or both.

B. Management

1. **Sensorineural hearing loss, or “nerve deafness,”** is managed in most cases with hearing aids. The type of hearing aid recommended will depend upon the severity of hearing loss as well as any physical deformity of the external ear. In addition to amplification, early intervention educational services may be recommended with emphasis on language acquisition in light of the hearing loss.
2. **Conductive hearing loss due to ear infection or effusion** will be managed by a physician, usually either the pediatrician or an otolaryngologist. Once the infection is appropriately treated, the hearing should return to normal. Conductive hearing loss due to anatomical abnormality of the mechanical structures of the ear may be managed by surgery, amplification, or a combination of the two.

C. Monitoring

1. **Sensorineural hearing loss.** Children with cleft lip/palate and sensorineural hearing loss should be tested every 4-6 months in order to assess any progression of hearing loss and to make adjust-

ments to amplification as needed for proper fit as the child grows.

2. Conductive hearing loss. In cases of conductive hearing loss periodic assessment will assist the managing physician by providing feedback regarding the efficacy of treatment in achieving and maintaining normal hearing status.

II. Genetics

The geneticist is responsible for identifying the etiology and/or pathogenesis of the cleft or craniofacial anomaly. The information is then used to discuss overall prognosis for the patient as well as recurrence risk for the parents, patient, and other family members. As with other birth defects, clefts and craniofacial disorders may be the result of chromosomal abnormalities, single gene disorders, and/or environmental factors/agents. Most commonly they are the result of multifactorial inheritance involving the interaction of an individual's genetic background with the environment.

Considerable progress has been made in the identification of causative factors over the past 10 years particularly in the area of single gene disorders. The genes responsible for several of the most well known genetically determined syndromes have been recently identified. However, at the time of this writing, molecular testing is infrequently utilized in clinical management. Working drafts of the human genome sequence have recently been published in *Nature and Science*. Several surprises have emerged. The number of human genes (roughly 30,000) is far less than originally estimated. Through a variety of genetic mechanisms including alternative splicing and regulation of transcription, the 30,000 genes code for an enormously complex array of proteins. Clearly, biology is no longer "one gene – one protein." It is now known that mutations in different genes may produce the same phenotype (e.g. FGFR1 and FGFR2 in Pfeiffer Syndrome). Different mutations in the same gene may result in different phenotypes (e.g. FGFR3 and achondroplasia, hypochondroplasia, thanatophoric dysplasia, and Crouzon Syndrome with acanthosis nigricans). The tissue distribution of a mutation may produce a range of phenotypes from a multisystem disorder to a tumor (e.g. GNAS1 and McCune-Albright Syndrome, fibrous dysplasia, and pituitary adenoma).

With respect to environmental factors, there are some agents, such as with the acne drug, Accutane, which are potent human teratogens with a high risk for craniofacial malformation in prenatally exposed fetuses, regardless of the infant or mother's genetic background. There are factors, such as cigarette

smoking, that increase the risk for cleft lip with or without cleft palate only in susceptible individuals. However, the genes that confer susceptibility to most cleft and craniofacial conditions remain to be elucidated. There is currently considerable interest in folic acid as a pre- or peri-conceptual treatment that might reduce the risk for cleft lip and palate as it does for spina bifida. Further study is needed to confirm early reports.

Three types of genetic mutations are under investigation in craniofacial disorders:

1. Those that increase an individual's susceptibility for a given error in morphogenesis but produce a phenotype only through interaction with other genes or environmental factors;
2. Those that produce phenotypes directly; and
3. Those that modify expression of disease producing genes and thus alter the phenotype.

Genetic advances are likely to improve the ability to diagnose and test for syndromes impacting craniofacial development. Understanding of the molecular pathogenesis of a condition will hopefully translate into novel strategies for treatment through manipulation of cellular pathways. Recognition of the factors impacting susceptibility and risk may lead to more effective strategies for prevention.

A. Cleft lip with or without cleft palate (CL±P)

1. Incidence in the general population is roughly 1:1000, but varies in different racial groups.
2. Although the majority of CL±P occurs in an otherwise normal individual, between 10% and 20% of affected individuals have the condition as part of a syndrome with broader implications to the individual and family. These conditions need to be identified such that appropriate follow-up is instituted and accurate recurrence risk counseling is offered.
 - a. The majority of syndromes are diagnosed clinically through history and physical examination.
 - b. Chromosomal testing may be indicated when CL±P occurs with other malformations, growth deficiency, or developmental delay.
 - c. Molecular (DNA) testing is available for a very few specific conditions.
3. For isolated CL±P, multifactorial inheritance is likely. Empiric risk for recurrence for unaffected parents with one affected child is 4:100 or 4%. This risk also applies to the affected individual's own chance for similarly affected offspring.
4. Prenatal diagnosis for isolated CL±P depends upon the ability of ultrasound to visualize the fetal face. For syndromes in which CL±P represents one feature, prenatal diagnosis should be

tailored to the underlying etiology of the syndrome.

B. Cleft palate alone (CP alone)

1. Incidence in the general population is roughly 1:2000.
2. Although the majority of CP alone occurs in an otherwise normal individual, up to 50% of affected individuals have the condition as part of a syndrome with broader implications to the individual and family. These conditions need to be identified such that appropriate follow-up is instituted and accurate recurrence risk counseling is offered.
 - a. The majority of syndromes are diagnosed clinically through history and physical examination.
 - b. Chromosomal testing may be indicated when CP occurs with other malformations, growth deficiency, or developmental delay.
 - c. Molecular (DNA) testing is available for a very few specific conditions.
 - d. Stickler syndrome is a common enough disorder that ophthalmologic evaluation of at-risk individuals is recommended.
3. For CP alone, multifactorial inheritance is likely. Empiric risk for recurrence for unaffected parents with one affected child is 3:100 or 3%. This risk also applies to the affected individual's own chance for similarly affected offspring. The risk is for an infant with CP alone, not CL±P.
4. Prenatal diagnosis for CP alone is currently not possible.

C. Non-cleft craniofacial abnormalities

1. This group of conditions is highly heterogeneous and runs the gamut from disorders of unknown etiology with a negligible recurrent risk (e.g., amnion rupture sequence) to those in which single gene mutations play the determining role (most of the syndromic craniosynostoses) with a substantial risk for recurrence in some families. Since prognosis and recurrence risk information is specific to each condition, genetic evaluation is encouraged in this population.
2. Prenatal diagnosis may be possible for a few conditions depending upon the etiology, the phenotype produced, and the availability of chromosomal and molecular diagnosis.

sists of assistance with infant feeding, access to team care, and family education. The nurse continues to interact with the family throughout all phases of the treatment period to assist them in understanding and complying with the recommended treatment plan, as well as providing crisis intervention and anticipatory guidance.

A. Prenatal

1. Assist with family education about clefting and other craniofacial disorders and team care after birth.
2. Provide information about potential feeding issues.
3. Introduction to Parent Support Network if applicable.
4. Provide direct contact information for team evaluation.

B. Neonatal

1. Initial contact with newborn in birth hospital – discussion of newborn care, team care, and early cleft management, feeding, resources, support group.
2. Modeling acceptance of child with craniofacial malformation.
3. Ongoing follow-up of feeding and weight gain after discharge, directly or through consultation with primary care physician.

C. Infant/Toddler

1. Preoperative preparation for surgical procedures, discharge teaching, and follow-up.
2. Ongoing coordination of team services/care.
3. Ongoing support of family.
4. Feeding issues – introduction of solid foods, prevention of bottle caries, weaning from the bottle, cup feeding.
5. Anticipatory guidance regarding growth and development issues; particularly encourage parenting techniques that promote speech development.

D. Preschool/School-Aged/Adolescents

1. Preoperative preparation that involves both the child and family.
2. Assistance with initiation of speech therapy and advocacy in the IEP process in obtaining services from the school district.
3. Ongoing evaluation of audiology and ENT concerns.
4. Referrals for social skills, self image concerns.
5. Introduction to other similarly affected patients/families.
6. Continued emphasis on multidisciplinary team care services.
7. Referral of adolescent/adult for genetic counseling.

III. Nursing

The role of nursing in the care of patients/families affected by craniofacial anomalies is multifaceted including education, case management, consultation, research, and primary care. Early intervention con-

IV. Oral and Maxillofacial Surgery

This discipline is concerned with the occlusion and facial form of patients with cleft and craniofacial anomalies. They work with other members of the team to ensure harmonious and appropriate dental arch form and facial form. Although there may be overlap with plastic surgery and otolaryngology and head and neck surgery in some areas, the oral and maxillofacial surgeon manages the alveolar cleft and skeletal problems related to cleft and craniofacial anomalies such as maxillary hypoplasia and other skeletal malocclusions.

- A. Prenatal: May counsel families regarding prenatal diagnosis and implications.
- B. Neonatal: May be involved in airway management. See Otolaryngology section for details.
- C. Infant: Early bone grafting of the cleft alveolus has been performed by some at this time but this is a controversial procedure that has been associated with poor mid-facial growth and class III dental malocclusion.
- D. Toddler: Occasionally primary teeth in the line of the alveolar cleft will need extraction at this age.
- E. Preschool: See Toddler section above.
- F. School-Aged
 - 1. Bone grafting of the alveolar cleft is usually done during the period of mixed dentition.
 - a. Age 6 to 10.
 - b. Orthodontic care prior to bone grafting to align the dental arches on either side of the cleft.
 - c. Sometimes teeth in and around the cleft can be salvaged with bone grafting saving the need for prosthetic dentistry later.
 - d. Depending on the size of the cleft in the alveolus the source of the bone graft may be the iliac crest, calvaria, or bone bank.
- G. Adolescents
 - 1. It is during this time when skeletal maturity is reached that consideration is given to maxillary or mandibular osteotomies to normalize occlusion and facial form.
 - 2. Patients with cleft lip and palate have a significant incidence of class III skeletal malocclusion with mid-face hypoplasia.
 - 3. This can be corrected with a Lefort I osteotomy of the maxilla sometimes combined with an osteotomy of the mandible. For slight anterior dental cross bites and orthodontic mid-facial protraction, a facial mask may be used.
 - 4. Distraction osteogenesis has been used to correct these skeletal occlusal problems as well.
 - 5. Skeletal surgery may be carried out in adulthood as well.

V. Orthodontics

Orthodontists are involved with the study and guidance of the growth and development of the face, and dentition of the child with a cleft or craniofacial anomaly from birth to maturity. Their role includes diagnosis of changing facial morphology and function due to treatment and growth. They provide orthodontic and orthopedic treatment and general expertise for consultation with all of the other members of the cleft and craniofacial team. Due to the long-term treatment required for the majority of these patients, different phases of active treatment, interspersed with periods of retention or no treatment, will be necessary.

- A. Prenatal – none
- B. Neonatal
 - 1. Pre-surgical infant orthopedics is sometimes used to reposition the segments of the cleft maxilla prior to lip repair. This can vary in complexity from lip taping to narrow the cleft, to a bonnet with elastic to ventroflex a protruding premaxilla, to more complex pinned appliances.
 - 2. These appliances can make lip closure easier. While this short-term benefit is clear, long term effects are unclear and controversial.
 - 3. Some clinicians use orthopedic appliances to alter the appearance of the nose and/or columella to improve the shape prior to lip repair.
- C. Infant

When the primary teeth begin to erupt, the parents are advised as to the possibility of dental irregularities, particularly an incisor or supernumerary tooth erupting into the palate. The long-term sequence of treatment is outlined in general terms.
- D. Toddler

No specific treatment is indicated, but digit habits and functional shifts may be addressed. Communication with the primary care dentist/pedodontist is established and future concerns outlined.
- E. Preschool
 - 1. In some cases, the maxilla may be expanded in order to improve dental function, eliminate functional shifts, to provide access for restorative care to carious teeth impacted in the cleft site, and/or to improve the nasal airway. However, long term retention is needed to maintain the expansion.
 - 2. Oronasal fistulae are sometimes a concern because of liquids escaping through the nose. The anterior part of the cleft may have become hidden as the maxillary segments moved together after lip repair, and this area may not have been repaired during palatoplasty. Consequently, palatal expansion may expose this oronasal communication. Surgical closure is often diffi-

cult, and the orthodontist may elect to use an obturator to close off the fistula.

3. A reverse pull headgear may be considered to protract the maxilla and maintain normal jaw relations. This is an effective treatment modality but requires considerable compliance on the part of the patient. Overall success is also uncertain due to the difficulty in anticipating future jaw growth when trying to compensate for inadequate maxillary growth.
4. Prior to pharyngeal flap surgery a lateral cephalograph is to be taken while vocalizing "Uu ..." and at Rest to evaluate the pharyngeal space architecture: length and width of the velum and the palatal height relative to the first cervical vertebral.*

F. School-Aged

1. Fixed appliance therapy usually occurs in the mixed dentition between the ages of 7 and 9 years, with the goal of preparing for alveolar bone grafting.
2. This phase usually involves aligning malpositioned incisors and expanding the maxillary arch to an appropriate relationship with the lower dental arch. When this is complete, an alveolar bone graft is placed and any oronasal fistulae closed. Maintenance of expansion with a palatal bar or removable appliance is required for some time since the grafted maxilla is unable to maintain the corrected arch form.
3. Reverse pull headgear therapy may be initiated or continued during this time period.

G. Adolescents

1. When the permanent teeth have erupted, definitive orthodontic treatment begins.
2. Treatment may involve surgical or orthopedic repositioning of the jaws to optimize jaw relations and occlusion. Close cooperation between the orthodontist, surgeon, prosthodontist (if necessary), and general dentist is required during this time.

H. Adults

Adults generally require the same treatment as children and adolescents with some possible exceptions. Since adults have completed growth, no possibility exists for influencing jaw growth through orthopedics. Additional or more extensive surgery may be required to achieve the same result. Alveolar bone grafts are less successful in adults, and thus may not be indicated if a graft would not carry significant benefits. Otherwise, a properly treated patient should have the same dental status

as a non-cleft person. All aesthetic and functional goals can and should be addressed.

I. Record keeping

This is an important part of the orthodontist's role on the cleft and craniofacial team, as it is necessary for assessment of treatment results.

1. Infant – Photographs should be taken regardless of any treatment. Casts should be made prior to and following any pre-surgical orthopedic treatment. Infant casts are important to assess the wide variability of cleft morphology and to compare the results of different treatments over time as growth occurs.
2. Preschool – Records taken during this time period depend upon treatment rendered. If palatal expansion is done, casts, photos, and a posteroanterior cephalogram are important to assess the result of treatment.
3. School aged – Full or orthodontic records should be taken prior to any orthodontic intervention, including incisor alignment and palatal expansion. These records should include, but are not limited to casts, photos, radiographs (panoramic, occlusal, periapical, and lateral/submentovertex/posteroanterior cephalograms), and clinical examination. Further, appropriate records, such as casts and photos, should be taken after treatment.
4. Adolescents – Full orthodontic records as above should be taken before and after definitive orthodontic treatment. Progress records should be taken before and after orthognathic surgery, and more often as necessary.
5. Adult – Full records should be taken as described above.

VI. Otolaryngology - Head and Neck Surgery

This is a surgical and medical discipline that is concerned with congenital malformations of the head and neck, and the problems associated with them. These problems include cleft lip and palate, breathing problems, feeding problems, hearing problems, and speech problems. There are areas of overlap with plastic surgery, oral and maxillofacial surgery, audiology, and speech-language pathology.

- A. Prenatal: The otolaryngologist is frequently called on to counsel parents with a prenatal diagnosis of a cleft or craniofacial abnormality. Counseling at this time includes information about potential early feeding and breathing problems, and should discuss timing of various surgical procedures. These include repair of cleft lip and palate, and placement of ear tubes.

* Being consider for addition to the Core Curriculum
– see Chapter 30.

B. Neonatal: During the neonatal period, the otolaryngologist should be involved with evaluating and assisting with adequate nutritional intake. Help is offered to the primary physicians with evaluating adequate weight gain. If there is poor weight gain, or difficulty with oral intake, then airway evaluation may need to take place. It is important to differentiate between primary feeding problems, and feeding problems secondary to airway problems. This includes sleep study, blood gases, and laryngoscopy and bronchoscopy.

Potential interventions for airway and feeding problems include insertion of nasal airways, tracheotomy, distraction osteogenesis of the mandible, and gastrostomy.

Hearing tests should be accomplished shortly after birth to obtain a baseline-hearing test. This should be done in the first 1-2 weeks of life before fluid accumulates in the middle ear, common to nearly all babies with cleft palate.

Ear tubes are placed on nearly all babies with cleft palate, and are usually done between 2 and 6 months of age. Hearing re-evaluation is done after the tubes are placed. Cleft lip repair typically is accomplished at 6-12 weeks of age, and palate repair at 6-18 months old.

C. Toddler

1. Monitor for development of obstructive sleep apnea.
2. Monitor for development of hearing loss.
3. Replacement of ear tubes, if necessary.
4. Prescribe hearing aids when appropriate.
5. Evaluate for cochlear implant when appropriate.
6. Monitor speech and advise beginning speech therapy.

D. Preschool

1. Continue hearing and speech monitoring.
2. Evaluate nasal airway for obstruction, and aesthetic appearance of nose.
3. Evaluate need for lip or palate revision.
4. Monitor for development of sleep apnea, and its cause.
5. Perform speech endoscopy and physical management of VPI if warranted.

E. School-Aged:

Continue to monitor and treat hearing, speech, and airway problems.

F. Adolescent and Adult

1. Perform septo-rhinoplasty if needed.
2. Perform tympanoplasty or other ear surgery if needed.

VII. Pediatric Dentistry

The role of pediatric dentistry in treating individuals with cleft and craniofacial anomalies is the comprehensive preventative and therapeutic oral health care of children from birth through adolescence and special patients beyond the age of adolescence who demonstrate mental, physical, and/or emotional problems. In addition, the pediatric dentist should provide preventative counseling and caries control to maintain the child's oral cavity in a state that maximizes the outcomes of therapies provided by other team members.

A. Prenatal

1. Parental information and support.
 - a. Maximize the families' support network.
 - b. Minimize the transmission of cariogenic bacteria from the parents to the child.
2. Provide information to parents about neonatal treatment options.
 - a. This will maximize their ability to make informed decisions about treatment options such as pre-surgical infant orthopedics.

B. Neonatal

1. Parental information and support.
2. Pre-surgical infant orthopedics (see the orthodontic section for more information).
3. Growth and development monitoring.
4. Caries prevention.

C. Infant

1. Caries prevention counseling.
2. Peri-operative care.
3. Infant orthopedics continued.
4. Growth and development monitoring.

D. Toddler

1. Caries prevention.
2. Growth and development monitoring.

E. Preschool

1. Caries prevention.
2. Growth and development monitoring.
3. Behavior modifications.
4. Routine dental care.
5. Interceptive orthodontics where appropriate.
6. Restorative procedures.

F. School-Aged: Same as preschool plus preparation for alveolar bone grafting where necessary.

1. Oral hygiene guidance.
2. Removal of primary dentition in surgical site.

G. Adolescents

1. Oral hygiene.
2. Periodontal concerns.
3. Support during comprehensive orthodontic treatment.
4. Caries prevention.

H. Adult

1. Preparation for transfer to general dentist or other dental specialist.
2. Monitor third molar and refer to OMFS where appropriate.

VIII. Plastic Surgery

Plastic surgery is the surgical discipline concerned with the restoration of normal form and function for patients with cleft and craniofacial anomalies. This is accomplished through appropriately timed operations throughout the patient's life. Some deformities can be reconstructed with one operation early in infancy and others require multiple surgical treatments as growth and development occur. There may be overlap with oral and maxillofacial surgery, and otolaryngology, head and neck surgery in the performance of these procedures. The goal is always to have normal function and appearance throughout a patient's life, realizing that this cannot always be accomplished because of anatomic or developmental considerations and how they relate to the timing of surgery.

A. Prenatal

1. Prenatal diagnosis of cleft and craniofacial anomalies is becoming more frequent with ultrasound.
2. Counseling regarding the implications and subsequent treatment may be carried out prior to birth.
3. Although fetal surgery has been done in animal models for cleft repair, this is not an accepted procedure for cleft repair at present.

B. Neonatal

1. Please see Section I for a detailed discussion of team evaluation and cleft classification.
2. Some surgeons are advocating cleft lip and palate repair in the neonatal period. The advantages of this approach have not been proven and the risk of complications is higher.

C. Infant: This is the typical time when surgical closure of the lip and palate is accomplished.

1. Cleft lip is usually surgically closed in the first 2 to 3 months of life when it is clear that the baby is healthy and thriving. Most surgeons still use the rule of tens to plan the timing of closure.
 - a. Ten weeks.
 - b. Ten pounds.
 - c. Hemoglobin of ten.
2. Some surgeons perform lip adhesion prior to definitive lip repair. This procedure is a partial lip repair that does not rearrange the structures into normal anatomic position. Its purpose is to narrow the cleft making the final lip repair easier.

3. The goal of the lip closure is to create a lip that functions well and approximates the physical characteristics associated with a non-cleft lip. The physical characteristics of the nose will also be improved by the lip closure. Sometimes lip revision will be required to improve the result, but the first operation generally provides a dramatic and lasting improvement in the function and appearance of the baby's lip.

4. The timing of palate closure varies from team to team but is usually carried out from 6 months to 18 months of age.
5. In children with airway problems or extremely wide palatal clefts, closure may be delayed.
6. The reason to close the palate is so that speech will develop normally and the patient will not regurgitate liquids and solids into the nose when eating.
7. Every sound in the English language except M, N, and NG resonates orally.
8. When a palate and cleft is not closed, resonance is hypernasal and multiple errors in speech development will occur.

D. Toddler

1. Despite closure of the palatal cleft, many children with cleft palate will still require speech therapy and approximately 10 to 20 percent may require secondary surgery for persistent hypernasal speech after closure.
 - a. This is called velopharyngeal insufficiency (VPI).
 - b. VPI becomes evident at age 2 to 3.
 - c. Rarely can occur without cleft palate.
 - d. Secondary surgical procedures to correct this problem are
 - 1) Posterior pharyngeal flap
 - 2) Pharyngoplasty
 - 3) Augmentation of the posterior pharyngeal wall
 - 4) Speech prosthesis
 - e. See Speech Language Pathology section for more details on evaluation.

E. Preschool

Nasal reconstruction may be performed just prior to kindergarten, possibly combined with a lip revision. These procedures are performed to improve function of the lip and nose, and to ensure that the child will look their best at a critical time of increased peer interaction when they begin school.

F. School-Aged

1. Dental concerns usually are primary during this time as orthodontics and alveolar cleft bone grafting are carried out.
2. See the Orthodontics and Oral and Maxillofacial sections.

G. Adolescents

1. Some children with clefts develop maxillary retrusion requiring jaw surgery to align their dental arches after their facial growth is complete (usually age 14 to 18).
2. After this is accomplished a final septorhinoplasty may be performed to improve breathing and nasal aesthetics.

H. Adult

1. Most patients have completed treatment by the time they reach adulthood.
2. Surgical revision is usually successful in treating any residual problems.

IX. Psychology and Clinical Social Work

The psychologist provides evaluation of, and treatment for, emotional, learning, developmental, and adjustment disorders. This generally occurs within the context of the patient's family. Particular attention is focused on the manifestations of appearance and speech on the patient's self esteem and coping strategies for the patient and family in dealing with issues related to multiple operations. The clinical social worker may also focus on many of these concerns as well as using their expertise to obtain services when needed for patients.

A. Prenatal

Assist with prenatal counseling regarding future expectations of development and coping with the unexpected intrauterine diagnosis of a child with a cleft or craniofacial anomaly.

B. Neonatal

1. May assess high risk infants for risk of developmental disorders.
2. May also assist parents with stresses related to children with facial deformity or other developmental problems.
3. Family support groups such as groups of parents of children with clefts or craniofacial anomalies can be very important to some parents in helping them cope with the birth of a child with a cleft or craniofacial anomaly. This can continue throughout childhood and adolescence.
4. In children with high risk for developmental problems, early referral to an infant program may be beneficial.

C. Infant

1. Infant assessment includes developmental assessment of motor and language development, and social responsiveness.
2. Continue to assess the family.

D. Toddler

1. Toddler assessment of self help skills, social development, and motor/language development.
2. Continue to assess the family.

E. Preschool Development

1. Evaluate language and intellectual development.
 - a. Expressive vs. Association language disorders are frequent and need to be carefully monitored.
 - b. Early verbal IQ deficits are common and may affect overall IQ scores.
2. Early Social Interactions.
 - a. Parent-child interactions.
 - b. Overprotectiveness may be present in parents of children with facial deformities and this should be monitored and counseling provided when needed.
3. Developmental Assessment.
 - a. Need for early assessment due to high frequency of early delay.
 - b. Validity problems of early assessment make it necessary to avoid rigid establishment of intellectual ability.
4. During the preschool years delays in development frequently first manifest themselves. The psychologist and speech language pathologist are the team members most likely to diagnose and recommend specific interventions to maximize the patient's potential development when delay is present.

F. School-Aged Child

1. Learning disorders in children with clefts and craniofacial anomalies.
 - a. Reading disorders
 - 1) Need for early screening intervention and remediation
 - 2) Reading problems related to speech problems are common
 - 3) Reading comprehension problems related to language problems may occur, thus reading evaluation should include assessment of both word recognition and reading comprehension
 - b. Memory disorders
 - 1) Late development of auditory memory
 - 2) Learning problems related to short term memory are frequent, therefore screening of short term memory or word finding problems is important (dysnomia)
 - c. Language disorders (common types)
 - 1) Dysnomia (word finding problem)
 - 2) Expressive Dysphasia (verbal expression of ideas problem)
 - 3) Associative Dysphasia (understanding of language problem)
 - 4) Behavioral Problems

- d. Acting out behaviors
 - 1) Related to early parent overprotection
 - 2) Related to language disorders
- e. Behavioral inhibition
 - 1) Anxious withdrawal
 - 2) Passivity (non-anxious) to avoid teasing
- 3. Teacher Expectations
 - a. Teacher perceptions of ability is often underestimated.
 - b. Self fulfilling academic expectations of teachers translates to underachievement.
- 4. It is during this time that children and their peers become aware of how they look. Deformities can lead to problems with teasing and self esteem in patients with clefts and craniofacial anomalies. The psychologist can help with coping strategies for patients and their families and can give the surgeon advice about the timing of operations for appearance during this critical time. The clinical social worker can aid in appropriate placement within the educational system.

Adolescents

- 1. Self-esteem
 - a. Realistic vs. unrealistic self perception of appearance and/or speech.
 - b. Social skills training may help to overcome hypersensitivity.
- 2. Behavioral Inhibition and Social Introversion
 - a. Depression/anxiety treatments may be indicated.
 - b. Social introversion as a way of life may result in lowered self expectation.
- 3. Dating and Self-Concerns
 - a. Cognitive behavior modifications may help through the use of self talk to provide strategies for coping with anxiety-provoking social situations.
 - b. Group counseling can be especially helpful with groups of peers with similar conditions.
- 4. During adolescence there is a heightened self-awareness of body image and greater existential worry about “who am I?”, “what is my identity”. Most adolescents experience these issues; however, the adolescent with facial differences or speech problems may experience a greater sense of “being different”, leading to greater emotional turmoil.

Also, adolescence is a period when there is often a decrease in open communication with parents and other adults. Therefore, it is important for the team psychologist, and/or social worker, to communicate and screen adolescents for possible emotional/social concerns. Monitoring of school achievements, peer activities, and social interactions may reveal when problems are occurring.

G. Adulthood

- 1. Social Adaptation
 - a. Marriage aspirations
 - b. Activities
- 2. Educational/Vocational Aspirations
 - a. Achievement motive
 - b. Work aspirations

X. Speech and Language Pathology

The speech/language pathologist provides evaluation and treatment of four communication parameters for patients with clefts and craniofacial anomalies, from infancy through adulthood. These parameters include resonance, articulation, phonation, and language development. The goal of the speech/language pathologist is to facilitate normal speech and language development. This is achieved by providing education concerning speech and language development, recommending and providing speech therapy, and as the child matures, by providing more direct perceptual, acoustic, sound pressure, radiologic, and aerodynamic measurements of the velopharyngeal mechanism. Dental, hearing, prosthetic, and surgical interventions must be factored into all management considerations. If velopharyngeal insufficiency is suspected, and palatal management is considered, direct visualization of the velopharyngeal mechanism during speech production is required, with repeat studies following surgical or prosthetic management.

A. Neonatal and Infancy

- 1. Monitor and assess feeding, swallowing, and hearing ability.
- 2. Discuss the following areas with family: language, cognition, and speech development with and without a cleft palate or palatal dysfunction.
- 3. Monitor and stimulate receptive and expressive language and cognitive development.
- 4. For babies with cleft palate, facilitate oral communication by emphasizing all vowel sound production and those consonants produced by the lips and anterior tongue, which are nasal, or require little intraoral air pressure (/m/, /n/, /w/, /l/, and “y”). Avoid consonant constrictions that are made in the back of the throat, in the glottal area, or made by the posterior tongue to posterior pharyngeal wall. Also, avoid excessive yelling and screaming.

B. Toddler (≤ 3 years)

- 1. Monitoring the patients’ general communication development, motor skills, and cognition.
- 2. By this age, patients have usually undergone lip and/or palate repair, and their speech, language, resonance, and voice needs to be assessed with consideration for early speech and/or language

therapy; with more global delays, an early childhood program should be instituted.

3. Nasal consonant substitutions may be observed. These occur when the speech articulators are placed appropriately for the intended oral consonant, but due to incomplete palatal closure, the speech sound is produced as a nasal consonant (/b/ becomes /m/; /d/ becomes /n/).
4. Compensatory substitutions may be noted. These are unconsciously learned speech patterns that occur when the articulators are positioned inappropriately in an effort to produce oral consonants. These are commonly heard in attempted production of *plosives* (sounds created by complete blockage of airflow followed by buildup of pressure which is suddenly released, such as /b/) and *fricatives* (sounds characterized by turbulent noise, such as /s/). If adequate oral pressure cannot be achieved with typical placement of the articulators, then an alternative constriction site may be used and pressure is created below the level of constriction. Some common compensatory articulations include:
 - a. Glottal stops – closure of the vocal folds at the level of the glottis.
 - b. Pharyngeal fricative – posterior positioning of tongue to posterior pharyngeal wall, occurring on fricatives and affricates.
 - c. Pharyngeal stop – posterior positioning of lingual base to pharyngeal wall, occurring on /k,g/.
 - d. Posterior nasal fricative – coarticulated nasal snort/flutter with any pressure consonant.
 - e. Mid-dorsum palatal stop – usually made in an approximate place of consonant /j/ in attempt to valve airflow.
5. Obturation of any hard palate fistulas may result in elimination of nasal leakage, improved resonance, and VP closure. Use of a speech bulb may be indicated for patients demonstrating reduced intraoral pressure, resulting in difficulty producing pressure consonants despite speech therapy.
6. Monitor phonation for vocal hoarseness, volume, and pitch levels with speech therapy for remediation or referral to otolaryngology.
- C. Preschool, School-Aged, and Adult: As speech articulation is acquired, the speech/language pathologist can begin differential diagnosis of velopharyngeal functioning.
 1. Continued monitoring of hearing acuity.
 2. Speech disorders related to velopharyngeal function.
 - a. Hypernasality – the perception of excessive nasal resonance during production of vowels and semi-vowels resulting from inadequate separation of the oral and nasal cavities.
 - b. Hyponasality – reduction of normal nasal resonance usually resulting from blockage of nasal airway by various causes.
 - c. Mixed hyper/hypo – simultaneous occurrence in the same speaker, usually resulting from incomplete velopharyngeal closure and high nasal resistance that is not sufficient to block nasal resonance completely.
 - d. Cul-de-sac – variation of hyponasality associated with tight anterior nasal constriction, often resulting in muffled quality.
 - e. Nasal air emission – nasal escape associated with production of high oral pressure consonants. Occurs when air is forced through incompletely closed velopharyngeal port, and can be audible or visible (evidenced by mirror fogging, nasal grimace, and/or nasal flaring).
 - f. Compensatory articulations.
 - g. Reduced intraoral pressure – reduced buildup of air in the oral cavity during production of pressure consonants due to inadequate valving of the VP mechanism.
3. Phonation: Voice quality, the perceptual characteristics of voice.
 - a. Hoarseness – a periodic vibration of the vocal folds producing a “rough” vocal quality.
 - b. Breathiness – excessive leakage of air through the glottis during phonation.
 - c. Pitch – sound property determined by the frequency of vibration of the vocal folds, either high, optimal, or low.
 - d. Volume – acoustic power or intensity.
4. Assessment
 - a. Perceptual
 - 1) Standardized articulation testing
 - 2) Assessment of perceived oral-nasal resonance balance during connected speech
 - b. Nasometer – nasalance, which provides a numeric output indicating the relative amount of nasal acoustic energy.
 - c. Aerodynamic Measurements – pressure flow studies estimating the sectional area of VP orifice (i.e., PERCI).
 - d. Assessment of oral structure and function.
 - 1) Face: symmetrical structure and function; drooling
 - 2) Lips: degree of bilabial contact, non-speech function, position during quiet breathing
 - 3) Dentition: occlusion, crossbite, open/closed bite, over/underbite, ectopic teeth, missing, rotated, or supernumerary, dental arch collapse, dental appliances

- 4) Tongue: deviation, lobule, frenulum, tongue thrust, non-speech function (range, strength, and symmetry of motion)
- 5) Hard palate: height, contour, width, oronasal fistulae
- 6) Tonsils/faucial pillars: size, position, and symmetry of tonsils, movement of pillars
- 7) Soft palate: symmetry at rest and during phonation; lateral and vertical degree of movement, uvula
- 8) Submucous cleft palate: bifid/notched uvula, zona pellucidum or transparency of the palate at midline, bony notching at the posterior border of the hard palate
- 9) Pharyngeal walls: vertical/lateral/symmetry of movement
- e. Imaging Studies

If VP dysfunction is suspected, direct visualization is required to evaluate velopharyngeal functioning during speech production using oral and nasal consonants in words, phrases, and sentences.

 - 1) Nasopharyngoscopy: degree of velopharyngeal closure for speech production and swallowing, velopharyngeal closure pattern, symmetry, velar contour, movement (velum, lateral pharyngeal and posterior pharyngeal walls, Passavant's ridge), adenoids/tonsils, laryngeal structure, and function
 - 2) Multiview videofluoroscopy
 - a. A midsagittal lateral view: movement of the velum and posterior pharyngeal walls, height and length of velum, point of velar closure, and velar relationship to adenoids and posterior pharyngeal wall; posterior tongue valving
 - b. Frontal view: lateral pharyngeal wall movement
 - c. Basal/Towne's view: all of the above, except vertical movement

The speech/language pathologist reviews both perceived speech characteristics and physiological status of the velopharyngeal mechanism during speech production, with possible recommendations for surgical or prosthetic management, speech therapy, and/or continued monitoring of VP function. If surgical management is recommended, perceptual evaluation should occur 3-6 months following surgery, with repeat imaging studies 6-12 months post management. Speech therapy for VP function should be deferred for 6-12 weeks following secondary palatal management, while therapy for developmental or compensatory articulations may be resumed in 3-4 weeks.

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14 Palatal Wound Healing: The Effects of Scarring on Growth

JOHANNES W. VON DEN HOFF, JAAP C. MALTHA, ANNE MARIE KUIJPERS-JAGTMAN

14.1 Introduction

Cleft palate patients often develop growth disturbances of the midfacial region after primary surgery. Factors such as intrinsic developmental deficiencies and functional distortions are initially involved but palatal repair seems to be a main factor in these growth disturbances [64–70]. A strong indication for the involvement of iatrogenic factors is the largely undisturbed maxillary growth in untreated patients [20, 46, 59]. In treated patients, the healing of surgical wounds originating from palatal repair is probably responsible for the growth disturbances [38]. This chapter gives an overview of the wound healing process with emphasis on wound contraction and scar formation since they are considered to be key events. Some specific features of the palatal wound healing process are highlighted. Further, the effects of palatal repair on growth of the maxilla and development of the dentition are reviewed, as well as possible means to improve the clinical outcome. This review is based on

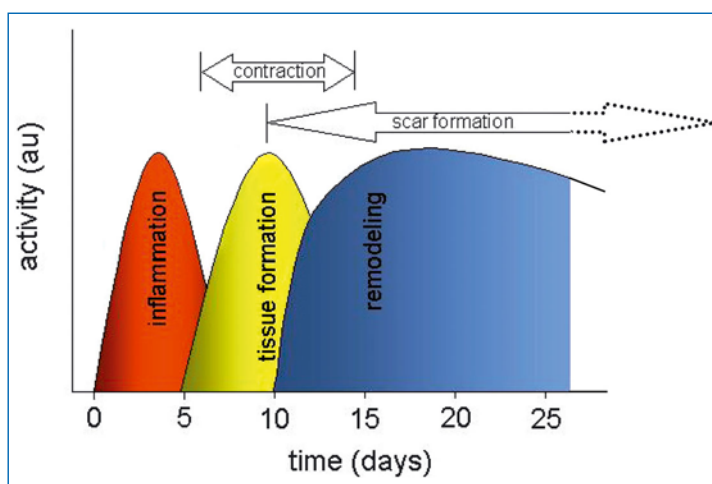
clinical evaluations, experimental research in animal models, and on in vitro experiments using cell culturing and tissue engineering techniques.

14.2 Wound Healing

14.2.1 Skin and Oral Mucosa

The general function of wound healing is to restore the integrity and function of the tissue. In tissues like skin and oral mucosa, wound healing involves a partly overlapping sequence of inflammation, tissue formation and tissue remodeling (Fig. 14.1). During inflammation, hemostasis is restored and bacteria and debris are removed from the wound. Subsequently, the defect is closed by the formation of new tissues and by wound contraction. Finally, tissue remodeling takes place during maturation of the newly formed tissues, which generally leads to the formation of a scar.

Fig. 14.1. Phases in wound healing. The wound healing process can be divided into three partly overlapping phases. Wound contraction is an early phenomenon while scar formation is a late phenomenon in wound healing (arrows)



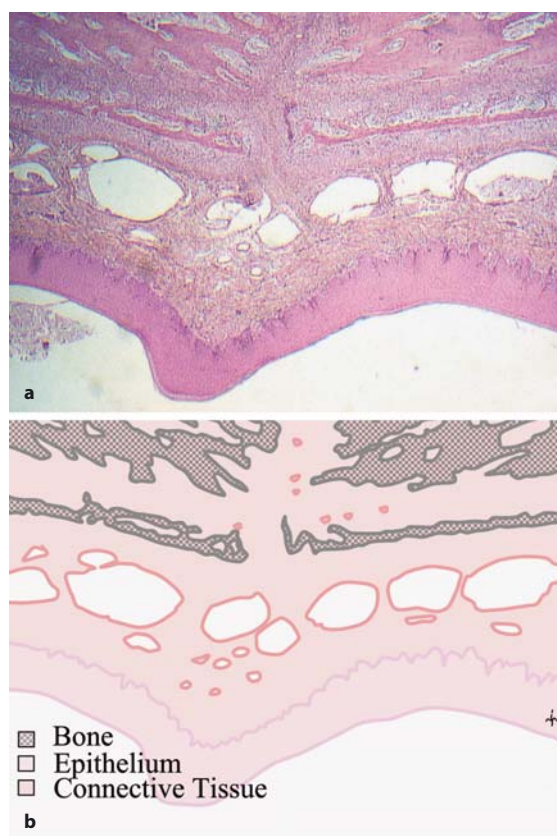


Fig. 14.2 a, b. The palatal mucoperiosteum. The mucoperiosteum of the palate contains an epithelium and a submucosal connective tissue, and is firmly attached to the palatal bone

In intraoral wounds, the wound healing process is generally faster than in skin and generates less scar tissue. Therefore, intraoral wounds are sometimes considered to be more similar to fetal wounds [57]. This may be related to the presence of lower levels of pro-inflammatory and pro-fibrotic cytokines in mucosal wounds [84]. The intraoral wound healing process is also influenced by the presence of saliva and large numbers of bacteria [94]. Saliva contains many growth factors such as epidermal growth factor (EGF). In addition, phenotypic differences between skin and mucosal fibroblasts may be involved [99]. These considerations, however, mainly apply to buccal mucosa, which has a quite different morphology than the palatal mucosa (Fig. 14.2).

In contrast to buccal mucosa, the palatal mucosa is a mucoperiosteum, which means that mucosa and periosteum are merged and attached to the palatal bone [81]. The palatal mucoperiosteum is also much stiffer than buccal mucosa and it contains less elastin fibers, as was also shown for the gingival mucoperiosteum

[9]. Furthermore, the epithelium of palatal mucosa is generally thicker than in the buccal areas, and it is cornified. All this implies that the physiological and mechanical characteristics of this tissue are quite different from buccal mucosa, which might explain differences in the outcome of the wound healing process.

Nevertheless, the general outline of both the palatal and the buccal wound healing process is similar to that of skin, which is described below [13]. It is important to stress that the phases of wound healing described here are not discrete episodes but they overlap in time (Fig. 14.1). In addition, the progress of healing in the outer wound area is more advanced than in the center, which means that subsequent phases of the wound healing process may be found in adjacent areas. This, of course, concerns only open wounds with a tissue defect in which healing takes place by second intention.

14.2.2 Phases in Wound Healing

Tissue injury causes disruption of blood vessels, and bleeding. Within seconds the coagulation cascade starts, leading to the formation of a fibrin-rich blood clot that contains numerous platelets. These platelets are reservoirs of cytokines and growth factors such as transforming growth factors (TGFs) and platelet-derived growth factor (PDGF) that attract inflammatory cells, especially neutrophils and macrophages. The blood clot forms a provisional matrix for the migration of those cells. Proteins such as fibronectin, fibrinogen, and vitronectin allow cell attachment and migration by their interaction with integrins, which are transmembrane cell surface receptors [91]. Neutrophils and macrophages subsequently clear the wound bed of debris and bacteria. In addition, the relative blood volume at the site of injury increases by dilation of the capillary vessels in the surrounding tissue and also by an increase of their permeability, which leads to redness and swelling. This phase is called *the inflammatory phase* and usually subsides several days after wounding (Fig. 14.1).

In the next phase, *the tissue formation phase*, keratinocytes, fibroblasts, and endothelial cells in the wound edge start to proliferate. They migrate into the wound bed, and start to form the neo-epithelium and the underlying granulation tissue [13]. This phase already starts a few days after wounding, before the inflammatory phase has come to an end. Keratinocytes seem to be activated by the partial loss of cell-cell contacts at the wound edge, and by locally produced growth factors such as epidermal growth factors (EGFs) and fibroblast growth factors (FGFs). Fibroblasts, endothelial cells, and more macrophages migrate into the wound bed. Their migration and other

activities are regulated by complex interactions with growth factors and extracellular matrix components within the provisional matrix [91]. Again, integrins play a major role in the interaction between cells and the extracellular matrix. The binding of matrix proteins to integrins is required for attachment and migration, and it also leads to the transmission of additional regulatory signals into the cells [63]. Cell migration furthermore requires the activity of matrix metalloproteinases (MMPs), enzymes that pave the way for cells by cleaving extracellular matrix proteins [49].

During migration into the wound, fibroblasts gradually switch to a more synthetic phenotype, a switch that involves the action of TGF β . The fibroblasts start to produce large quantities of collagen, with collagen type III as the main species, but elastin is not synthesized in the wound. Within this matrix, endothelial cells form numerous capillaries to nourish the newly formed tissue. This process is called neo-angiogenesis and FGFs, vascular endothelial growth factor (VEGF), and many other growth factors are implicated in its regulation as shown by in vitro studies [13]. As soon as the wound is filled with granulation tissue and the neo-epidermis is formed, the collagen production is reduced, which requires interferon- γ (IFN γ). A negative feedback mechanism based on the accumulated collagen may also contribute to the decrease in collagen synthesis. This event marks the beginning of the third and last phase, *the remodeling phase*.

The remodeling phase starts within 1 week after wounding and will ultimately lead to the formation of scar tissue. The remodeling of the extracellular matrix is mainly carried out by fibroblasts. It involves the degradation of collagen type III by matrix metalloproteinases [49] and the concurrent deposition of type I collagen by fibroblasts. In the second week after injury, fibroblasts also start to produce proteoglycans. The mechanical properties of the tissue are not only determined by collagens, but to a large extent also by these proteoglycans, since they can bind large amounts of water. In addition, many proteoglycans have been shown to regulate cell function, either by direct modulation of cell adhesion and proliferation, or indirectly through the binding or release of growth factors [53].

At the start of the remodeling phase, part of the fibroblasts within the granulation tissue differentiates into myofibroblasts that possess contractile properties. These specialized cells are strongly involved in the process of wound contraction. Their differentiation seems to be governed mainly by mechanical tension within the matrix, by TGF β , and by a specific variant of fibronectin, the ED-A fibronectin [85]. Wound contraction causes a rapid reduction of the surface area of the wound and a concomitant re-

arrangement of the collagen fibers. In the mean time the neo-epidermis is maturing into a fully differentiated stratified epithelium. After 1–2 weeks, no further contraction takes place because the myofibroblasts have disappeared, probably by apoptosis [22]. The induction of apoptosis is not completely understood, but several genes are known to govern the process. The expression of these genes is regulated by growth factors as well as by changes in the interaction between the cells and their extracellular matrix. In the next several months many of the fibroblasts, but also endothelial cells, disappear by apoptosis, which gradually renders the tissue less vascularized and less cell rich. The slow remodeling of the collagen fibers by remaining fibroblasts, which is part of the transition to scar tissue, can go on for a long time.

14.2.3 Contraction and Scarring

Wound contraction and scarring seem to be the two main processes in wound healing responsible for the growth disturbances after cleft palate repair, and are therefore reviewed here in more detail. Wound contraction is the reduction of the wound surface area by approximation of the wound edges and it may account for up to 80%–90% of wound closure [47]. The evolutionary function of this feature obviously is to speed up wound closure and thereby reduce the risk of infection and dehydration. Scar formation might be a negative side effect of this primarily beneficial process.

The cause of wound contraction is not yet exactly known. Two main theories have been described in literature. The first one states that fibroblasts, which migrate from the wound margins into the wound bed, cause traction in the extracellular matrix. This tensional force would be sufficient to contract the wound [24]. This theory does not require specialized cells to explain wound contraction. The second theory assumes that a specialized subtype of fibroblasts, the myofibroblast, is responsible for wound contraction [21, 27]. During wound contraction, fibroblasts containing intracellular stress-fibers are found within the granulation tissue. These stress-fibers have been shown to contain alpha-smooth muscle actin (ASMA), a cytoskeletal protein also present in smooth muscle cells. This protein seems to be required for the contraction of myofibroblasts within the granulation tissue. The coordinated contraction of myofibroblasts, attached to the extracellular matrix, causes the reduction of the wound surface.

Nowadays, the two theories have merged into a consensus theory stating that both fibroblasts and myofibroblasts are involved in wound contraction [85]. Initially, migrating fibroblasts in the wound area

generate tension within the matrix. The resulting strain within the matrix triggers the differentiation of fibroblasts into myofibroblasts, which also requires the presence of TGF β_1 . The coordinated action of myofibroblasts strongly increases tension within the wound tissue, which subsequently contracts. Thus, in the consensus theory, both fibroblasts and myofibroblasts contribute to wound contraction.

During contraction of the granulation tissue, extensive collagen remodeling takes place in which MMPs play a prominent role [49]. As a consequence of this remodeling, collagen type III is gradually replaced by collagen type I. The new collagen is deposited in an orientation that is guided by the main lines of tension within the extracellular matrix [71, 31]. The reorientation of collagen fibers and the substitution of type I collagen for type III collagen mark the start of scar tissue formation. If a uniform direction of tension exists within the contracting granulation tissue, the new collagen fibers will also be deposited in a uniform orientation. Consequently, the resulting tissue will develop the properties of a scar, a process that may slowly progress for several months to years [71].

During scar tissue formation, the number of endothelial cells and fibroblasts within the developing scar tissue slowly decreases, a process in which apoptosis is involved. The final scar tissue therefore is poorly vascularized and has a low cell density. In addition, elastin fibers, which provide elasticity to normal mucosa and skin, are not resynthesized during wound healing and scar tissue formation. Their absence and the presence of highly oriented collagen fibers make the scar a rigid and stiff tissue. A specific feature of palatal wound healing is the attachment of the scar tissue to the palatal bone (see Sect. 4.1). This may cause palatal repair to have considerable effects on maxillary growth.

14.3 Effects of Palatal Repair on Growth

Apart from embryonic distortions and intrinsic growth deficiencies, facial growth in cleft lip and palate patients may be affected by surgical repair, orthodontic treatment, and functional adaptations [38, 64–70, 72, 76]. Since the landmark studies of Graber [49] and Dahl [19], numerous descriptive cephalometric studies have been published (for an overview, see [76]). It is reasonably well established that cleft surgery, in particular lip and palate repair, can disturb normal growth and development of the maxilla in cleft patients [7, 38]. However, the possible growth effects of surgery should be evaluated in relation to the intrinsic abnormalities of craniofacial growth in cleft palate patients. This requires that unoperated cleft lip

and palate patients should be studied as well [11, 20, 39, 46].

Of all surgical procedures that are used in cleft lip and palate patients, palatal surgery has attracted the greatest amount of attention. The reason is that during this procedure mucoperiosteal flaps are created on the palate to close the cleft, leaving areas of denuded bone. The scar tissue that is formed during healing might be a potential inhibitor of subsequent maxillary growth and dental arch development. Many studies over the past 50 years have focused on the effects of specific techniques of primary palate repair on midfacial growth and development. The effects of palatal closure seem to be mainly confined to the maxillary base and arch [77, 38]. The maxilla is shown to be narrower, shorter, and displaced posteriorly relative to the cranial base. The dentoalveolar processes are often deflected to the median, resulting in anterior and transverse cross bites. However, since both lip and palatal surgery are generally performed, it is difficult to distinguish between the effects of the two types of surgery.

A problem with the evaluation of the literature on this subject is that the majority of the publications suffer from major methodological drawbacks, which minimizes their value [38, 69]. Hardly any studies are available that directly compare two types of treatment in a prospective research design. In contrast, there is a vast amount of retrospective studies available that all attempted to relate specific maxillary growth effects to particular surgical procedures. The most comprehensive study of this type is the multicenter study by Ross [64–70]. By comparing lateral cephalometric radiographs collected from 15 cleft palate centers from around the world, they concluded that it was difficult to isolate the effects of individual palate repair techniques. However, an inhibition of anterior growth and translation of the maxilla was a common finding. Another problem that is only addressed by few, is that not only the surgical technique but in particular the skills of the surgeon are very important for the long-term outcome in terms of growth and development [38, 78].

In conclusion, strong consensus exists that primary surgery is a major factor in the impairment of dentomaxillary growth. The extent of growth impairment may be influenced by the specific techniques, the timing and sequence of operations, the use of orthopedic appliances, and possibly the most important of all, the skills of the surgeon. No particular technique has been shown to produce consistently better growth results than any other. Assuming that scar tissue is a primary etiological factor in maxillofacial growth disturbances, most contemporary repair techniques attempt to minimize scarring. Animal experiments are very well suited to determine the exact effects of specific surgical procedures. For a better understanding of the

biological mechanisms in wound healing, and for the goal-directed modulation of healing, experimental studies are of major importance. Results of such studies are discussed in the next section.

14.4 Experimental Research

Extensive research in animal models has been performed to evaluate the effects of cleft surgery on growth and development of the maxilla, and to study the wound healing process. Animal models are also used to develop new surgical techniques that may reduce the unfavorable effects of surgery. Tissue engineered constructs are being developed to prevent attachment of scar tissue to the bone, or as a substitute for the lacking mucosal tissue. A lot of *in vitro* research is also aimed at the elucidation of aspects of the oral wound healing process.

14.4.1 Effects of Surgery on Growth

Since suitable animal models for congenital clefts are not available, the effects of reconstructive surgery are evaluated with surgically created clefts in dogs, rabbits, and rats. Two different approaches have been used to evaluate the effects of surgery on midfacial growth and development of the dentition. The first approach is to create a cleft in the soft tissue and the palatal bone by surgical means. This cleft is subsequently closed again as the actual experimental intervention. It is obvious, however, that a surgically created cleft is different from a congenital one. Such a cleft creates a surgical trauma that might act as a confounding factor for the interpretation of the results. Only if the bony cleft is considered to be essential for evoking the disturbances in growth and development does this approach make sense. Bardach and coworkers have used this model since 1975. They performed some of the earliest experiments on the possible negative effect of lip repair [1–6]. Lip repair in rabbits and beagle dogs with surgically created complete unilateral clefts, was found to result in a significant increase in lip pressure and a corresponding maxillary growth deficiency. The authors therefore suggested a causal relationship between the two.

The second approach is based on the assumption that the soft tissue intervention is crucial for the growth disturbances after cleft palate surgery. Already in the late 1960s Kremenak and coworkers performed mucoperiosteal excisions without affecting the palatal bone in young beagle dogs as a model for the clinical situation after cleft palate repair. This approach led to growth disturbances that were similar to those after surgical cleft closure in children. Kremenak therefore

concluded that “in this model, mucoperiosteal denudation of palatal shelf bone adjacent to deciduous molars is the single surgical variable responsible for the maxillary growth disturbances seen” [37]. This is in agreement with later dog studies in which a mid-palatal soft tissue cleft was created that was subsequently closed by von Langenbeck’s technique (e.g., [88, 89]). Their results not only show the same effects on growth, but also that the extent of this effect is related to the age at which surgery is performed. Growth disturbances turned out to be most prominent when surgery is performed before shedding of the deciduous dentition. Furthermore, these studies, as well as recent studies in rats [36], show that the deviations in maxillary arch dimensions are not only caused by a decreased sutural growth but also by a palatal tipping of the teeth in the lateral areas. In dogs, this tipping is especially prominent when surgery is performed at a young age, and it becomes apparent only after shedding of the deciduous dentition.

An explanation for this effect can be found in the healing of the soft tissue wounds and more specifically in wound contraction and scar tissue formation. Wound contraction in dog mucoperiosteum is most prominent in the first week after surgery. In palatal wounds in rats it has been shown that the number of myofibroblasts increases considerably in that period [17], which also seems to be the case in dogs. Thereafter, the maturing granulation tissue is characterized by a gradual decrease in the number of fibroblasts and inflammatory cells, and an increase in number and thickness of collagen type I fibers [74]. Elastic fibers are not present in the granulation tissue, or in the scar tissue at later stages.

A specific feature of the healing of open wounds in the mucoperiosteum is the deposition of callus-like cancellous bone on the palate. The granulation tissue adjacent to the palatal bone acquires an osteogenic potential, or osteogenic cells migrate into that area from adjacent periosteal tissues, and new bone is formed. This phenomenon is also known from other craniofacial bones and from long bones, where removal or mobilization of the periosteum leads to callus formation. Most of the collagen fibers of the scar are oriented in a transverse direction, but many fibers also show a vertical orientation. These vertical fibers become embedded in the cancellous palatal bone as Sharpey’s fibers, generating a strong attachment of the scar tissue to the underlying palatal bone (Fig. 14.3).

The transverse fibers appear to be continuous with the cervical periodontal ligament, thus forming a mechanical connection between the teeth and the mucoperiosteal scar tissue [89, 36]. At the end of the growth period, the teeth alongside the scar tissue show a palatal tipping which is probably caused by the

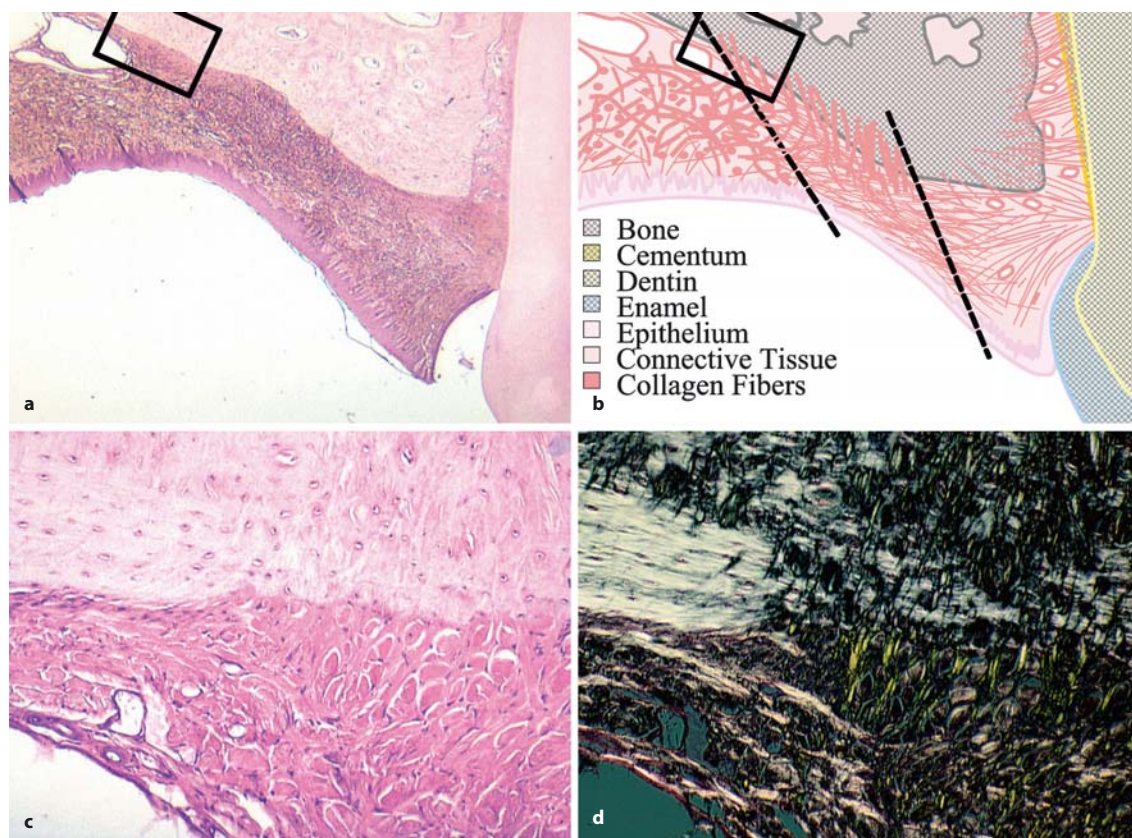


Fig. 14.3 a–d. Palatal scar tissue. **a** shows the scar tissue adjacent to the teeth. The organization of the tissue is shown schematically in **b**. **c** and **d** shows an enlargement of the *squared*

section. Note the thick perpendicular collagen fibers (Sharpey's fibers) running in to the bone, which are more clear when seen in polarized light (**d**)

traction of the scar tissue on the erupting permanent dentition. These findings have led to the hypothesis that the iatrogenic effects of palatal surgery are initially caused by wound contraction, but scar tissue formation and the accompanying attachment of the scar tissue to the palatal bone and the teeth are probably the most important features. This leads to a restriction of maxillary growth and to a palatal tipping of erupting teeth in that region.

14.4.2 Modification of Surgical Techniques

Several researchers have tried to modify cleft palate surgery in order to avoid the appearance of denuded bone areas and the subsequent growth impairment. Perko [60] was the first to use a mobilized mucosal split flap for palatal closure. The disadvantage of his technique was the high risk of necrosis because the flap was only pedicled at the dorsal side. This technique has been modified by Leenstra et al. to obtain a

flap that is pedicled both at the dorsal and the ventral side [41, 42]. They used a partially split flap, leaving the lateral bone covered with the osteogenic layer of the mucoperiosteum, without impairment of the major neurovascular bundles [42]. In dogs this technique was promising, since it led to less attachment of the mucoperiosteum to the underlying bone, and an improved transversal growth and development of the dentition [42, 43], and also in a clinical setting the results were promising [41].

The denuded bone can also be covered with a bio-material, which can either be applied as such or supplemented with cultured cells. This type of approach belongs to the field of tissue engineering.

14.4.3 Tissue Engineering

Tissue engineering has been defined in the late 1980s as “the application of principles and methods of engineering and life sciences toward fundamental under-

standing of structure-function relationships in normal and pathological mammalian tissues and the development of biological substitutes to restore, maintain or improve tissue functions” [79].

With respect to cleft palate surgery a variety of approaches have been chosen to improve the outcome of the wound healing process. Firstly, biocompatible membranes have been used to prevent attachment of the scar tissue to the palatal bone or to reduce contraction and scar formation. A second approach is the engineering of mucosal substitutes to replenish the tissue defects. To this end, thin layers of keratinocytes have been cultured that can be used as epithelial grafts. Alternatively, keratinocytes have been cultured on top of a dermal substrate to produce a bilayered or composite graft, which is a substitute for the entire mucosa. The dermal substrate may consist of a collagenous matrix without cells or a matrix with cultured fibroblasts.

14.4.3.1 Biocompatible Membranes

Biocompatible synthetic membranes have been used to inhibit the attachment of scar tissue to the palatal bone by covering the denuded bone areas after surgery. Initially, the principles of guided tissue regeneration were used by inserting membranes in the mucoperiosteal defects [33]. These membranes were supposed to cover the palatal bone, inhibit osteogenic processes and thereby prevent the formation of Sharpey's fibers. Bio-resorbable poly-(L-) lactic acid membranes and nonresorbable polymer membranes yielded unsatisfactory results. This was caused by an uncontrollable degradation of the lactic acid membranes, and exfoliation or incomplete coverage of the bone by the nonresorbable membranes [45].

Next to synthetic membranes, collagen-based membranes have been used for intraoral surgery. Atecollagen membranes have been successfully used to improve gingival healing in a rat model [50]. Similar membranes were used later in a model for cleft palate repair in young rabbits [26]. In a split-mouth design, the membranes were implanted on the denuded palatal bone at the experimental side, while the control side was left open. The authors report that implantation reduced contraction, and allowed more favorable growth of the palatal bone and normal development of the dentition.

To further improve collagen-based membranes, suitable growth factors might be added to promote angiogenesis and regeneration of the submucosa and the overlying epithelium [103].

14.4.3.2 Epithelial Sheets

Oral keratinocytes, obtained from a mucosal biopsy, can be cultured to form an epithelial sheet. The keratinocytes are generally expanded using the Rheinwald and Green method or a modification thereof [62]. Grafts cultured from autologous keratinocytes seem to behave as a permanent epithelial substitute after transplantation [86, 8]. If allogeneic keratinocytes are used, however, the graft behaves as a temporary wound dressing that only accelerates re-epithelialization [83]. Similar results were obtained after skin grafting with cultured epidermal keratinocytes. Several studies indicate that wound contraction and scar formation may still occur after the application of a cultured epidermal graft [14, 90]. In addition, the epithelial sheets are very fragile and difficult to handle.

14.4.3.3 Composite Substitutes

An approach that is more widely used nowadays was developed for the grafting of full-thickness burn wounds on the skin. In this approach, the epithelium is cultured on a dermal substrate to produce a composite graft [61]. The presence of a dermal substrate is supposed to diminish contraction and subsequent scar formation. It can be derived from human dermis or prepared from purified collagen and additional extracellular matrix components. Also, fibroblasts and growth factors may be included within the dermal substrate to improve vascularization and epithelial differentiation. An overview of the production of a composite cultured graft is given in Fig. 14.4.

If dog palatal keratinocytes are cultured on skin-derived substrates they form an epithelium similar to that in vivo [58] (Fig. 14.5). Such a composite graft containing human keratinocytes showed a good clinical take rate after intraoral grafting. Compared to a substrate without cells, the composite graft showed enhanced epithelialization, and maturation of the submucosa [34]. Grafts composed of a collagen substrate and human keratinocytes inside the matrix have been implanted in full-thickness skin wounds in immunodeficient mice [10]. They showed a reduced wound contraction and a stimulation of epithelial maturation.

Composite grafts containing both keratinocytes and fibroblasts have also been constructed. The fibroblasts within the dermal substrate seem to enhance the differentiation of the overlying epithelium. Preliminary studies have shown that keratinocytes

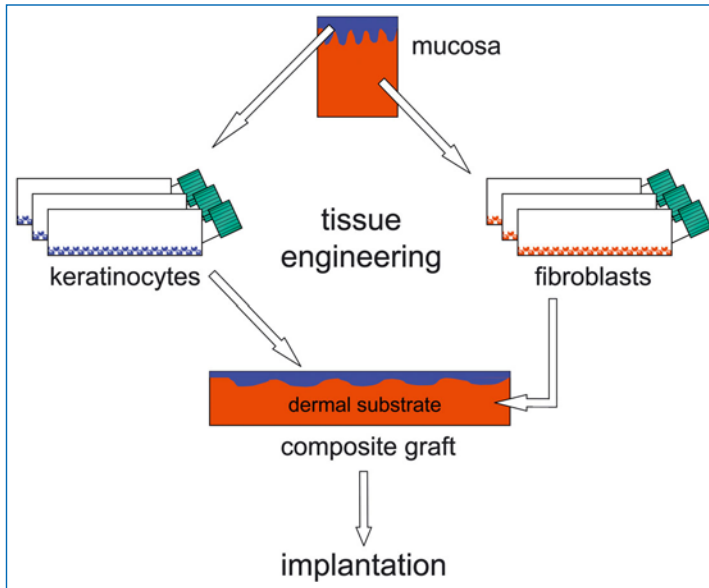


Fig. 14.4. Tissue engineering of palatal mucosa. A mucosal biopsy is taken and divided into epithelium and submucosa. Keratinocytes are cultured from the epithelium and fibroblasts from the submucosa. The fibroblasts are seeded into a dermal substrate and the keratinocytes are cultured on top to construct a composite graft for implantation

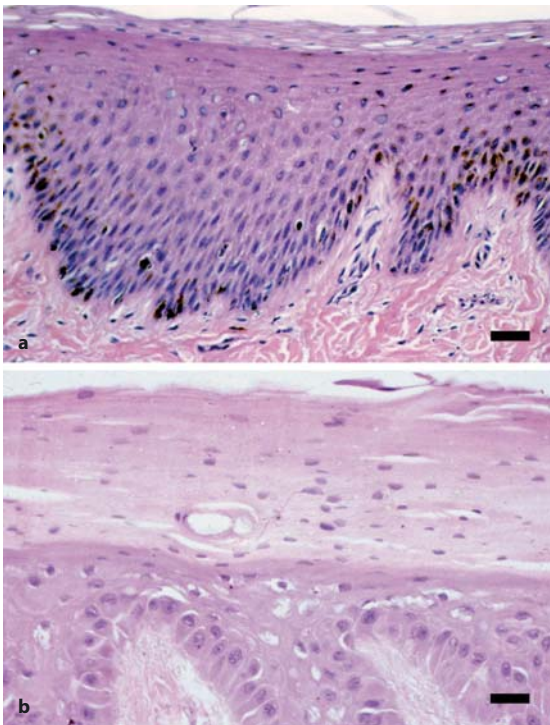


Fig. 14.5 a, b. Tissue engineered mucosa. Dog palatal keratinocytes were cultured on a de-epidermized dermis (DED) for 3 weeks. **a** shows normal dog palatal mucoperiosteum. **b** shows the cultured graft. Note that the cultured graft has a much thicker cornified layer due to the absence of mechanical abrasion

cultured on a collagen gel containing fibroblasts form a well-differentiated epithelium [32]. Transplantation of such a graft into skin wounds in immunodeficient mice reduced wound contraction [52]. Up to now, composite grafts have not been evaluated extensively for intraoral transplantation. However, their suggested capacity to reduce wound contraction and subsequent scarring makes them good candidates for application in cleft palate surgery. The use of tissue engineering techniques may well lead to the development of suitable grafts for the improvement of cleft palate repair.

14.4.4 Mechanisms of Wound Healing (In Vivo Studies)

Recent animal experiments on cleft palate surgery focus mainly on the biological processes during intraoral wound healing and the possibilities of reducing contraction and subsequent scarring. Observational research aims at a more detailed description of the cytokines and growth factors involved in the intraoral wound healing process. Most of these studies have been performed in rats. Several proinflammatory factors such as interleukins (ILs) are involved in palatal wound healing. For example, IL-1 seems to be essential for intraoral wound healing but not for dermal wound healing. Its effects are probably mediated by an increase of the antibacterial activity of PMNs and monocytes [30]. On the other hand, oral wounds contain less IL-6 than skin wounds, while the expression of IL-10 in both types of wounds is similar [84].

TGF β s and FGFs are also very important in the early phases of wound healing, as they are involved in the differentiation of myofibroblasts. TGF β_1 is supposed to upregulate FGF receptor-1 and FGF receptor-2 on palatal fibroblasts and thus increasing the susceptibility of these cells to FGFs. The latter factors are mainly produced by macrophage-like cells and cause an increase in the number of myofibroblasts [25, 35, 93]. On the other hand, IFN γ inhibits the differentiation of myofibroblasts from rat palatal fibroblasts in vitro [93] and in vivo [16]. FGF2 is also involved in the re-epithelialization of palatal mucosa since a single topical application already speeds up this process [56]. After the completion of wound contraction and re-epithelialization, myofibroblasts disappear, probably through apoptosis that again is influenced by TGF β_1 and FGF2 [25]. The same processes have been described for skin wounds, but they seem to proceed at a slower pace [55].

In the later phases of palatal wound healing the number of cells and the amount of collagen is higher than in the normal mucoperiosteum. The collagen type I fibers are densely packed and transversely aligned, indicating a distinct scar tissue [15] with a decreased vascular density [12]. Since IFNs possess antifibrotic and anticontractive properties, they have been used for experimental research on skin wound contraction and the reduction of keloids. IFN α inhibits wound contraction [54], IFN β downregulates collagen synthesis of dermal fibroblasts in vitro [23], and IFN γ downregulates collagen synthesis in skin wounds [29].

Strong evidence for the involvement of growth factors in the regulation of wound contraction and scar formation has been obtained from studies on fetal wound healing. Intrauterine wound healing in mammalian fetuses occurs without contraction and scar formation up to a certain gestational age [101]. In this early phase of gestation, myofibroblasts do not occur during wound healing and the regenerated tissue is indistinguishable from normal tissue. This scarless healing seems to depend on a specific growth factor profile within the wound and on the typical intrauterine environment. Several researchers have utilized these characteristics of fetal wound healing to perform prenatal cleft palate surgery in animals [94, 98, 105]. These studies are debatable since they involve only surgically created clefts.

Recently, a model for congenital cleft palate in goats was described in which clefting was induced by the teratogen anabasine [106]. In affected fetuses, palatoplasty was performed in utero using a modified von Langenbeck technique at 85 days' gestation. At 6 months of age, the mucoperiosteum of the palate had healed without scarring, and the function of the soft palate and the architecture of the velum were sim-

ilar to that of unclefted controls. However, it is highly unlikely that prenatal surgery for cleft palate repair will soon be common practice because of diagnostic and ethical problems [102]. The effects of specific growth factors that are possibly involved in scarless fetal healing can be further investigated by in vitro studies.

14.4.5 Mechanisms of Wound Healing (In Vitro Studies)

Several cell culture models are exploited to investigate aspects of fibroblast and keratinocyte biology that are relevant to the oral wound healing process. These studies often aim to elucidate the differences between oral and dermal wound healing or to find pharmacological means to modify the oral wound healing process. Two-dimensional monolayer cultures are suitable to study the expression of certain proteins or the proliferation and migration of cells. However, to study the interactions of cells with their extracellular matrix, a three-dimensional culture model is required. An additional advantage of three-dimensional culture models is that the physiology of the cells is more similar to that in vivo [51].

In two-dimensional monolayer cultures, human oral keratinocytes constitutively express higher levels of hepatocyte growth factor and keratinocyte growth factor than skin keratinocytes [57]. In addition they express higher levels of IL-6 after stimulation with other cytokines [100]. These differences may contribute to the preferential healing of mucosal wounds in comparison with skin wounds. For rat palatal wound fibroblasts, it was shown that interferons can reduce their collagen synthesis, which seems to be favorable in the light of scar reduction [15]. Oral fibroblasts have also been shown to possess a lower migration capacity in vitro compared with skin fibroblasts [99]. Fibroblasts, taken from subsequent phases in rat palatal wound healing, show distinct expression patterns of integrins and cytoskeletal proteins [87]. This indicates that specific subpopulations of fibroblasts may be responsible for wound contraction and scarring.

Fibroblasts cultured in three-dimensional collagen lattices contract this lattice in a time-dependent way [96]. The contraction is caused by attachment of the cells to the collagen and their migration through the lattice. This model therefore represents some aspects of the wound contraction process in vivo. Several investigators have used this model to compare the contraction capacity of oral and dermal fibroblasts [40, 97, 104]. In general, these studies show that oral fibroblasts have a higher contraction capacity than dermal fibroblasts, similar to that of fetal fibroblasts

[97]. The contraction of all three types of fibroblasts is inhibited by IL-1 β . Fibroblast-populated collagen lattices were also used to investigate the migration of fibroblasts into a wound [95]. To this end an experimental wound was made in the lattice, and the migration of fibroblasts into the wound was measured. Wound repopulation by mucosal fibroblasts was greater than that by age-matched dermal fibroblasts. These in vitro models contribute to the understanding of cellular processes during oral wound healing and the identification of factors with potential therapeutic value. Eventually, this may lead to new strategies to reduce intraoral wound contraction and scarring, and subsequent growth disturbances.

14.5 Application of Experimental Results

The available data from clinical as well as experimental research clearly indicate that primary palatal surgery is an important factor in the maxillary growth disturbances in cleft palate patients. Experimental research in animal models shows that wound contraction and subsequent scarring in the surgical wounds are key events. More specifically, the attachment of the scar tissue to the palatal bone and the periodontal fiber system inhibits sutural growth and normal development of the dentition. This can only be partially prevented by modifications of the surgical technique such as the split flap technique, which avoids the denudation of palatal bone. Another development is the use of tissue engineering techniques to construct a substitute for oral mucosa, which can be used to cover the denuded bone areas. There are some indications that this may reduce contraction and subsequent scarring on the palate. Whether this approach will yield better growth parameters after palatal surgery remains to be established. A third approach might be the pharmacological reduction of contraction and scarring after palatal surgery. In vitro studies have shown that some cytokines have the capacity to inhibit specific aspects of scar formation such as the differentiation of myofibroblasts and excessive collagen deposition. Only few of these factors have also shown favorable effects in vivo. However, the efficient delivery of the factor into the wound and the targeting to specific cells are practical problems that have to be solved.

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15 Lip and Palate Surgery

SAMUEL BERKOWITZ

15.1 The Influence of Surgery on Growth

The anteroposterior and vertical growth of the midface is not yet completely understood. The stimulus or growth force is still being debated. However, it is essential to understand the basic maxillary growth concepts to appreciate the influence of palatal and facial surgery on midfacial growth.

Enlow [1] states that:

Growth is not “programmed” within the calcified part of the bone itself. The “blueprint” for the design, construction, and growth of a bone lies in the functional matrix: the composite of the muscles, tongue, lips, cheeks, integument, mucosa, connective tissue, nerves, blood vessel, airways, pharynx, the brain as an organ mass, tonsils, adenoids, and so on. Growth fields (growth sites) for example, a suture and the alveolar bone housing teeth, throughout a bone do not have the same rate of growth activity. Some “fields” grow much more rapidly or to a greater extent than others. The same is true for resorptive fields. All surfaces are sites of growth; relocation of the bone, going from one location to another, is the basis for remodeling.

In the maxilla, the palate grows downward (that is, becomes relocated inferiorly) by periosteal resorption on the nasal side and periosteal deposition on the oral side. The nasal mucosa provides the periosteum on one side and the oral mucosa provides it on the other side; together growth remodeling is paced by the composite of soft tissue housing the palate. This results in a downward relocation of the whole palate-maxillary arch composite.

The bone tissue that comes to house the teeth at older age periods is not the same actual bone enclos-

ing them during the succession of former growth levels.

Depending on the extent of palatal surgery and the resulting scar tissue, the stability of a region can be disrupted if the results negatively affect the pattern of resorptive and depository fields on bone and at the suture; that is, scarring can work against growth. As the midface grows, bone is laid down in the sutures surrounding the maxillary complex. Any damage to one of these sutures can interfere with the direction and amount of growth. For example, an excessive amount of scarring at the pterygomaxillary suture (PTM) or at the premaxillary vomerine suture (PVS), will interfere with anteroposterior and vertical maxillary and premaxillary growth (Fig. 15.1).

Growth movement of the premaxilla is produced by the growth expansion of all the bones behind and above it and by growth in other parts of the maxilla, especially at the premaxillary vomerine suture (PVS). The premaxilla itself contributes a major part of its own forward growth movement through changes at the PVS. These displacement growth movements are a result of the “carry effect,” as Enlow calls it, which is produced by the expansion of the soft tissues associated with the bones, not a “pushing effect” of bones against bones. Scarring of the palatal mucoperiosteum, therefore, acts to interfere with the “carry effect,” thus preventing the change in position of the maxilla within the face.

Berkowitz speculates that pressure forces created by the uniting of the cleft orbicularis oris are directed posteriorly through the maxilla and nasal septum to the maxillary sutures and premaxillary vomerine suture, respectively. This action is similar to that of a head gear (cervical traction) used to control midfacial growth in orthodontics. These forces interfere with the ability of the premaxillary vomerine suture and the pterygomaxillary suture to function properly



Fig. 15.1. Complete bilateral cleft lip and palate cast (*top*) and occlusal x-ray film (*bottom*). → (*bottom*) indicates premaxillary vomerine suture (PVS) at *a* (*bottom*)

and lead to some degree of midfacial retrusion. Berkowitz's clinical findings support the concept that maxillary growth can be restrained by a variety of force systems. Marked deformation and gross changes can result when large forces are used over long periods of time. Maxillary maldevelopment is three-dimensional, resulting in a decrease in midfacial height, length, and width.

15.2 Surgical Closure of the Cleft Lip and Palate

Enthusiasm for a particular form of therapy should not be regarded as scientifically established when, in fact, it has not been subjected to critical scientific analysis. Treatment fads come and go. Unfortunately, in cleft palate surgery, it takes at least a decade to determine the effectiveness of a procedure. A prior belief in a particular therapy often determines the biased selection of evidence to support a concept currently in vogue; some clinicians show a select sample of cases to prove the theory only in part, if at all.

The following chapters will present our understanding of the cleft defect and the face in which it exists. These chapters are designed to answer some basic questions: What is the natural history of the cleft defect? How do similarly classified clefts differ from each other? What should be done for a child with a cleft lip and palate, and when should it be done? And finally, how does the treatment vary from child to child?

Although selected cases will be shown to develop our treatment philosophy, it must be stressed at the start that the concepts being presented are supported by findings from many longitudinal facial growth studies already published by our clinic and others world-wide. The cases from our center are unique to the extent that they represent the results of only one surgeon who is considered a master at his craft. Treatment failures as well as successes will be presented to develop and stress the physiological principles that are the basis of this treatment philosophy.

Improvements in surgical and orthodontic treatment methods, coupled with a better understanding of the natural history of cleft palate growth and development, have led to different yet frequently successful treatment protocols. Although some readers may be concerned that variations in successful treatment strategies interfere with comparisons and eventual standardization of treatment outcomes, nothing could be further from the truth. The one conclusion that should be drawn from these reports of clinical innovations is that there is more than one successful treatment protocol.

If the sample populations are large enough, any statistical evaluation of a generally successful treatment approach will show a certain proportion of failures. Obviously, a number of different physiological surgical procedures can be successful, provided that other dependent variables, such as the facial growth pattern and geometric form of the cleft defect, are complementary.

Fig. 15.2 a, b. Incomplete bilateral cleft lip and palate. **a** Presurgery. **b** 1 year, 6 months (1-6) after the lip is united. The prolabium extends to the vermilion borders



There is no reason to think that, by standardizing treatment protocols (as to type and timing), the same surgical procedure invariably will be successful. Certainly the skill employed is equally important. Moreover, it is a well-known fact that the same surgery, performed by the same surgeon on the same cleft type, can yield different results. Why? To find an answer to this question it is necessary to change one's primary focus from solely the surgery employed to the cleft defect itself and the face in which it exists. Doing so enables the clinician to take into account three mutually dependent critical variables: (1) the cleft defect; (2) the facial growth pattern and (3) the surgical procedure (see Chap. 16, Changing Philosophy of Surgery of the Cleft Lip and Palate in Goteborg, Sweden).

15.3 Lip Surgery

In a comprehensive evaluation of maxillary growth and development, the principles and techniques of tissue repair are of paramount importance as an integral part of any maxillary surgery. The basic requirement of any soft tissue closure, whether in unilateral or bilateral clefts of the lip, is that the full width of the lip be maintained. Clinical observations have shown that a moderately tight lip will hamper, to some extent, maxillary growth and produce some deformity with subsequent malocclusion of the permanent dentition. Skoog [2] has written that, in the cleft deformity, there is a variable but absolute tissue shortage. He concludes that, to be effective and achieve an anatomically correct and aesthetically pleasing result, reconstruction must therefore preserve and utilize all available tissue. Millard [3] has stressed that, in bilateral clefts of the lip, the prolabium – regardless of its size – should always be positioned to the vermilion border (Fig. 15.2).

15.3.1 Lip Adhesion

The cleft lip can be closed at any time from the first day of life, but the rule of “over ten” is usually followed. At 10 weeks of age, 10 pounds, and 10 g of hemoglobin, elective surgery is generally considered safe and the lip and nose have increased enough in size to facilitate the detailed surgery. Thus, most surgeons recommend surgical repair of the cleft lip at about 3 months of age.

Incisions in the sulcus and undermining of the soft tissues on the external surface of the maxillary segments were once commonly used to facilitate lip closure and decrease lip tension, especially in wide clefts (Fig. 15.3). Walker et al. [4], Bardach and Eisbach [5], Bardach and Klausner [6], Bardach et al. [7], and Collito [8] questioned the advisability of performing this procedure because they observed clinically that it contributed to secondary maxillofacial deformities and midfacial growth inhibition because of excessive scarring. This objection led Walker et al. [4] to present their concept of the Collito-Walker (C-W) technique of uniting the cleft lip (i.e., the use of lip adhesion without undermining; Figs. 15.4, 15.5). Bardach et al. [7], using rabbits, showed that lip repair with soft tissue undermining led to significant shortening of the maxillary complex. There is no reason to believe that the same negative effect would not also occur on children with lip/alveolar clefts, and therefore Bardach et al. [7] concluded that the lip adhesion procedure is preferred even though the posterior-directed pressure of the united lip potentially can slow down midfacial growth.

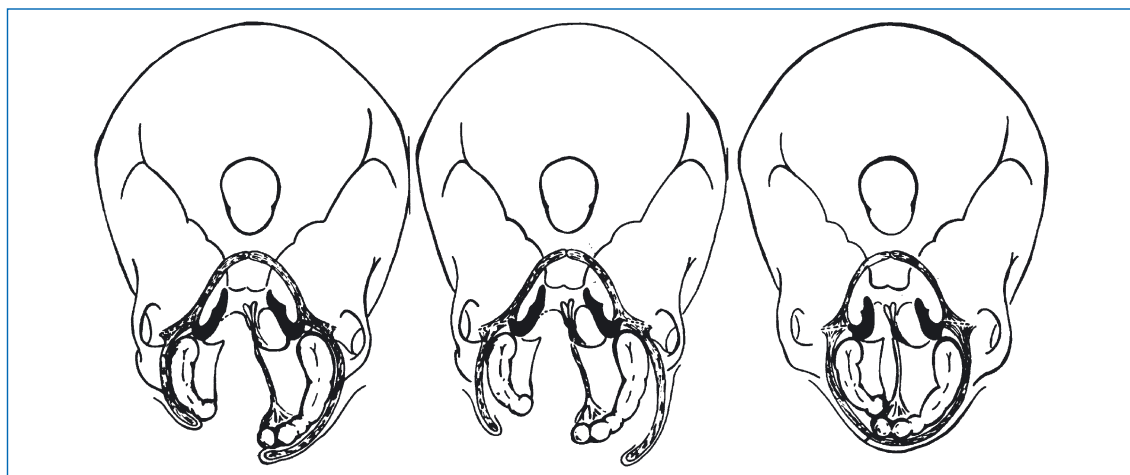


Fig. 15.3. Surgery to unite the lip over a large cleft space in one procedure. Undermining the soft tissue on the buccal alveolar surface posteriorly to the maxillary tuberosity prior to uniting the cleft lip has led to scarring and maxillary deformity. This

surgery is no longer being performed and has been replaced with a lip adhesion or the use of external elastics as the first procedure of choice. (Courtesy of [58])



Fig. 15.4a-d. Lip adhesion. **a** Complete unilateral cleft lip and palate at birth. **b** Lip adhesion at 3 months. **c** After definitive lip repair at 7 months. **d** Symmetrical lip at 11 months



Fig. 15.5a-c. Lip adhesion in an incomplete bilateral cleft lip and palate. **a** At birth. **b** At 3 years, after lip adhesion and definitive lip surgery at 8 months. **c** Full face at 3 years, showing good “cupid’s bow” and lip and nose symmetry

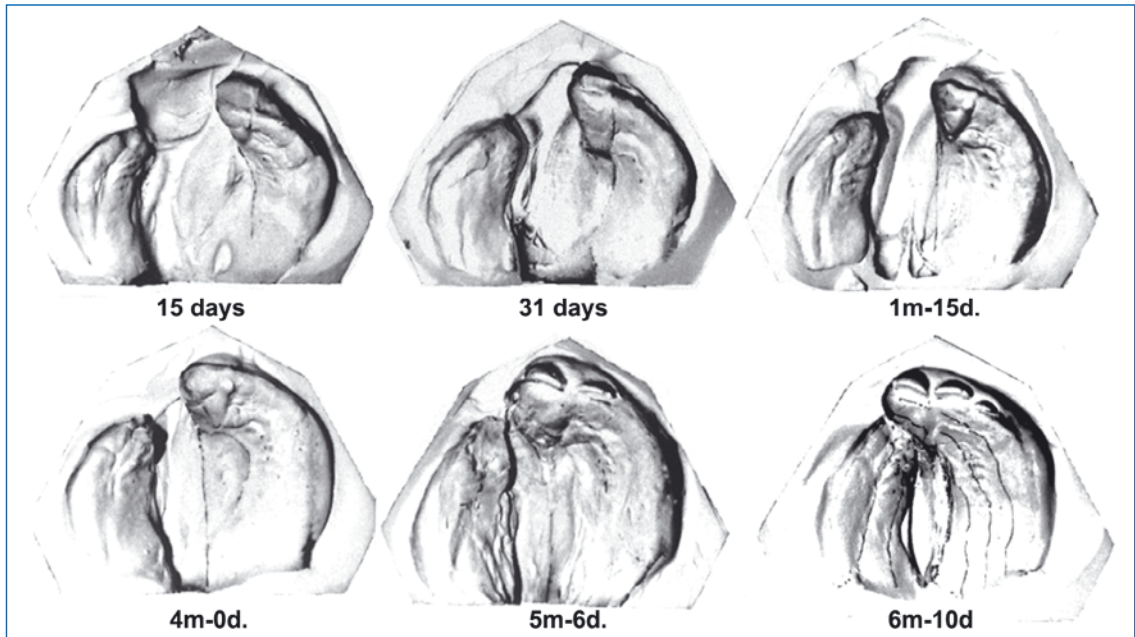


Fig. 15.6. An example of MOLDING of overexpanded lateral palatal segments. In CUCPLP a lip adhesion was performed at 3 months of age without prior presurgical orthopedics. The alveolar cleft gradually closed with the alveolar segments overlapping

15.4 Palatal Cleft Surgery: Type, Timing, and Sequence (Figs. 15.6–15.8)

15.4.1 What to Do and When to Do It: Speech and Palatal Growth Considerations

15.4.1.1 False Premise 1/2 Wrong Conclusions 1/2 Therapeutic Folly

At the turn of the last century, surgeons involved in cleft palate treatment usually performed surgical procedures whose sole purpose was to “close the hole” as early as possible without considering the ultimate effect of the surgery on palatal, facial, or speech development. These procedures, Millard [3] reports, fall into three categories:

1. The use of various kinds of flaps from other parts of the body to cover the cleft space.
2. Treating the edges of the cleft so that they could be sutured together by pulling the mucoperiosteum over the cleft. Failure of a lasting union led to the use of laterally positioned relaxation incisions by Dieffenback in 1826 and von Langenbeck in 1862 [9] (Figs. 15.9–15.11).
3. Staged surgical treatment.

Harold Gillies [10], an early proponent of staged surgical treatment procedure, mistakenly believed that the maxillary and mandibular arches were in a normal occlusal relationship at birth. He did not appreciate the distorting effect of aberrant muscle forces on the geometric relationship of the palatal segments to each other. He did, however, suggest that the lip be united soon after birth, and that the soft palate be detached from the hard palate and “pushed back” to increase its length. The cleft in the hard palate was to be closed before speech generally began at 2 years of age. For some surgeons this is still the best surgical sequence to follow.

In the 1920s William Wardill [11] focused his interest on controlling air flow through the nose. He appreciated the value of the velum along with the levators as a part of the sphincteric muscle system. Because uniting the soft palate alone often did not bring good speech, many surgeons (e.g., [11–14]) believed, as Gillies [10] did, that the velar length was insufficient in the von Langenbeck procedure and advocated “pushback” procedures [15].

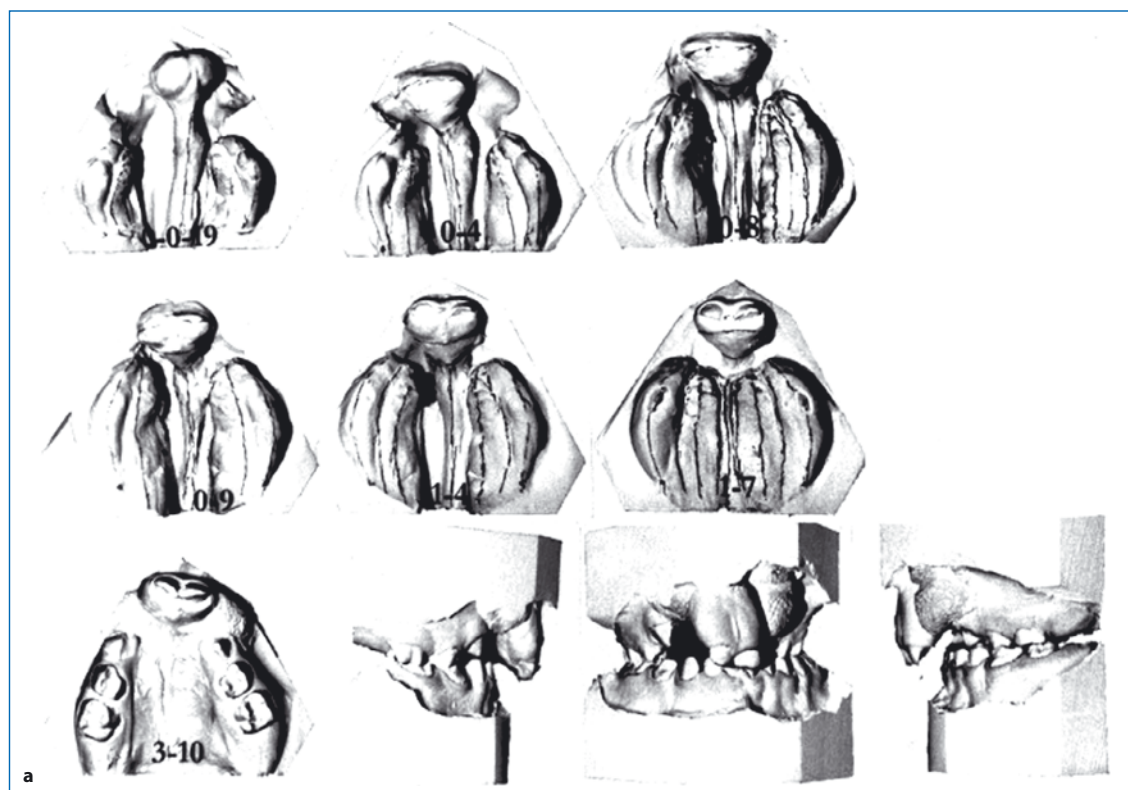


Fig. 15.7. a Lip adhesion causes premaxillary ventroflexion. Class II malocclusion with severe overjet. Anterior cleft space remains after 3 years, 10 months. No decision about correction

of the overjet should be made at this age. An obturator closing off the cleft space will improve speech and feeding. Palatal growth needs to be considered for future treatment

In 1937 Kilner [16] in London and Wardill [13] in New Castle, publishing independently, described a technique of palatal repair that ultimately came to be known as the V-Y retroposition operation. Wardill [13] and Kilner [16] both adopted the Veau technique for anterior repair, and the resulting “Veau-Wardill-Kilner” operation consisted of: (1) lateral relaxing incisions, (2) bilateral flaps based on posterior palatine arteries, (3) nasal mucosa closed as a separate layer, (4) fracture of the hamulus, (5) separate muscular layered closure, and (6) V-Y type of palatal lengthening (Figs. 15.9–15.12).

Soon various pharyngeal flap techniques were introduced when it was suspected that the velum was still too short even after it was moved back [17].

In 1958 Kilner [18] listed what he believed should be the aims of cleft palate treatment, in order of importance, as: speech, chewing, and aesthetics. This order of priorities is still preferred by many plastic surgeons, as well as speech-language pathologists. As a result, the surgical treatment concepts have been slow to change. To “close the hole as early as possible” with

or without velar pushback is still a widely prevailing concept.

“What to do and when to do it” were questions that had no universally accepted answers. Although surgeons such as Veau [11, 12] who appreciated the morphological differences in cleft types, still believed that closing the cleft palate early would improve speech development, many others began to think differently and emphasized the need for normal midfacial and palatal development. They recommended that cleft palate closure be postponed until either the deciduous or the permanent dentition had erupted. Advocates of delaying palatal closure were influenced by Graber [19, 20] and also by Slaughter and Brodie [21], who were disturbed by the results of Brophy’s [22] and other procedures in vogue at that time. They wanted to avoid secondary malformations to the palate and severe deformities of the maxilla caused by extensive mucoperiosteal undermining with wide lateral areas of denuded bone which led to severe scarring. Similar malformations were created in animals by Kremenak et al. [23, 24, 25].

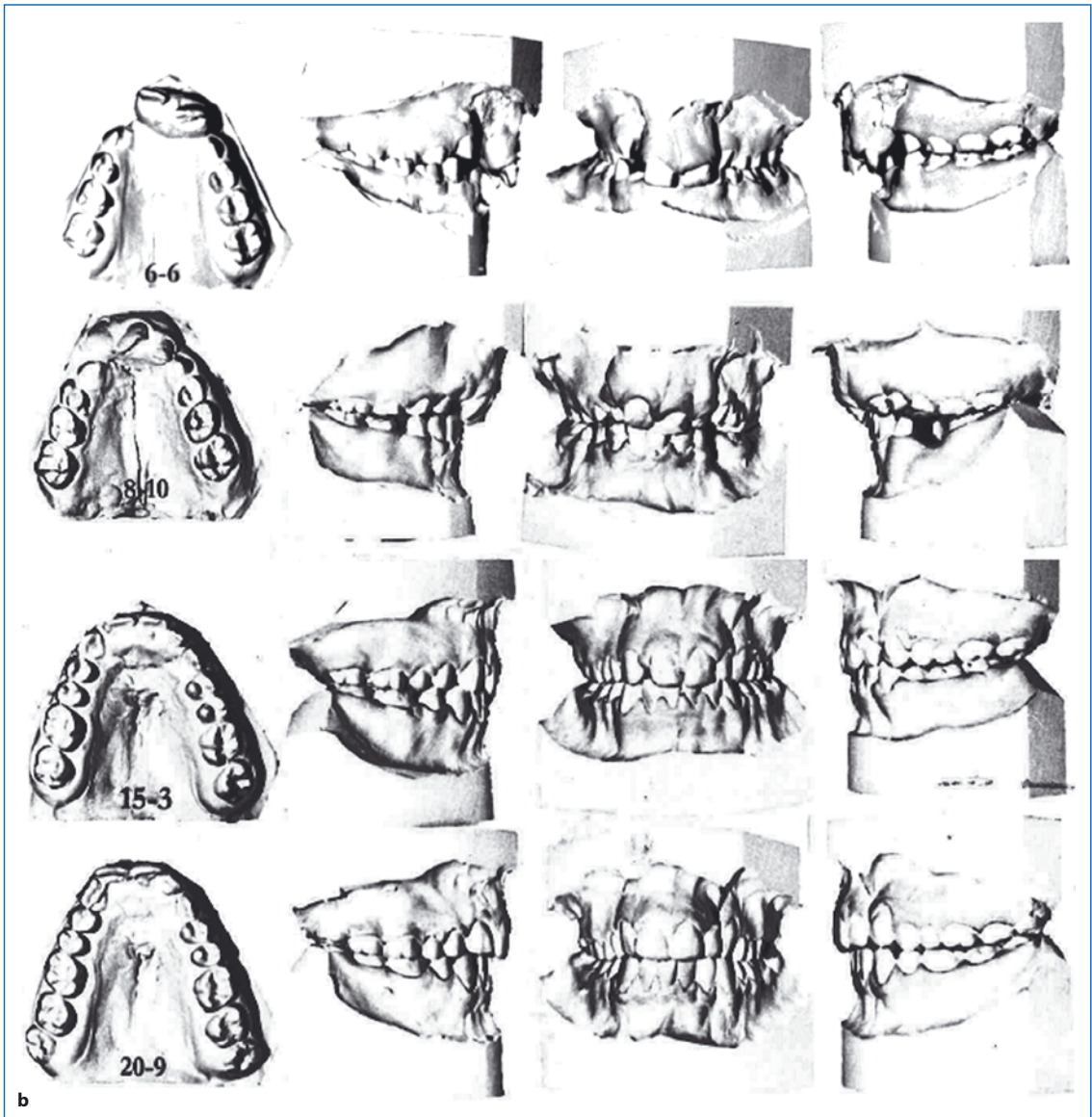


Fig. 15.7. b CBCLP. Serial cast of 2 cases (Fig. 15.7a,b and Fig. 15.8) demonstrate that the premaxilla and palatal shelves may react differently during the first years to the forces created by a lip strap attached to a head bonnet followed by lip surgery. In Fig. 15.7: Slow reduction of the anterior cleft space characterized this case. Although ventroflexion occurs, the anterior cleft space (the space between the premaxilla and the lateral palatal

segments) remained until 3 years and 10 months of age with a severe premaxillary overjet and overbite. No surgery to the premaxilla was necessary other than alveolar bone grafting at 8 years. The following series of casts show the attainment of excellent aesthetics, occlusion and speech. Palatal surgery using a von Langenbeck with vomer flap was performed at 23 months of age

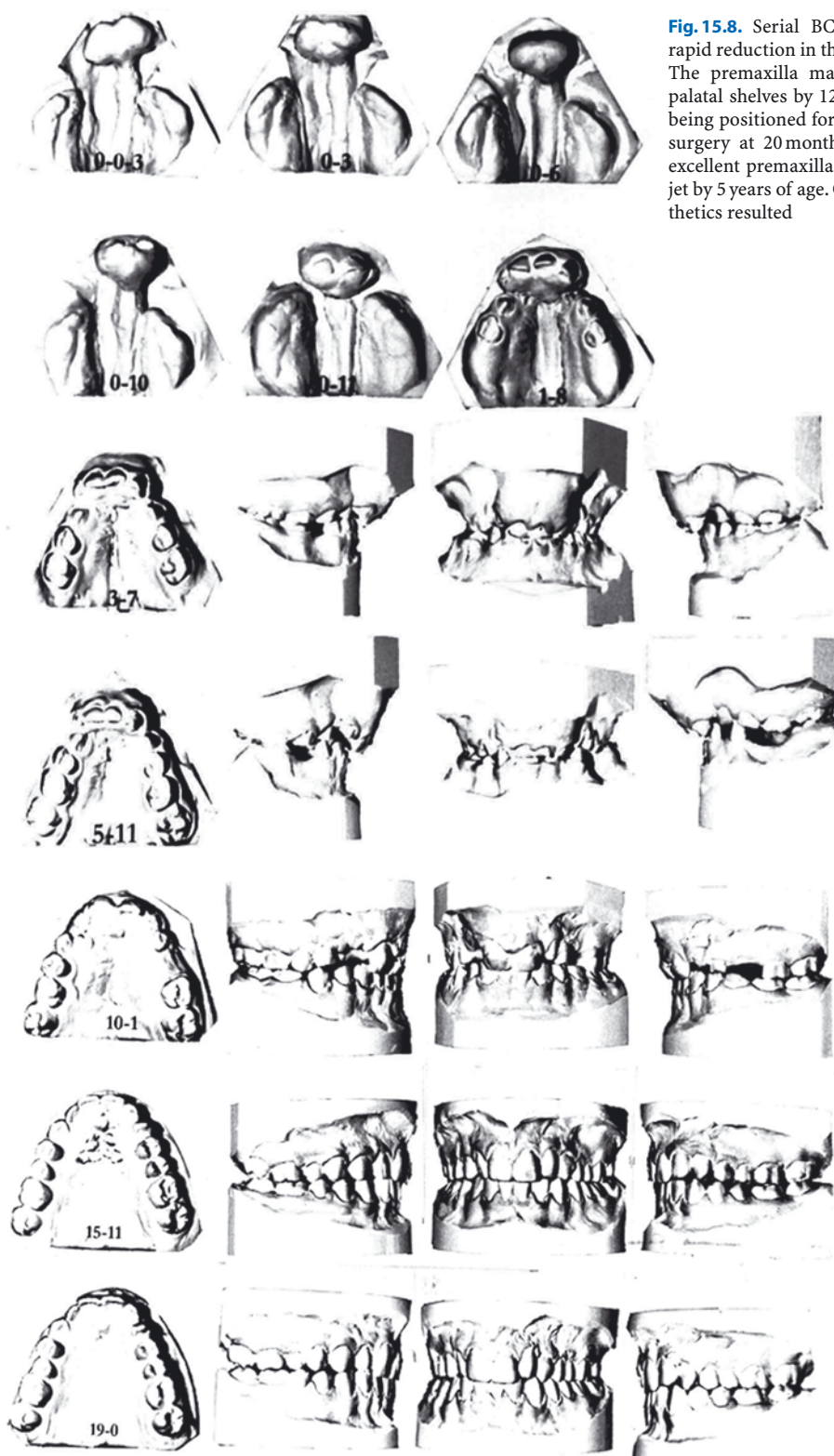


Fig. 15.8. Serial BCLP casts showing a rapid reduction in the anterior cleft space. The premaxilla made contact with the palatal shelves by 12 months of age while being positioned forward of them. Palatal surgery at 20 months of age resulted in excellent premaxillary overbite and overjet by 5 years of age. Good speech and aesthetics resulted

Fig. 15.9. von Langenbeck (simple closure) palatoplasty for isolated cleft palate

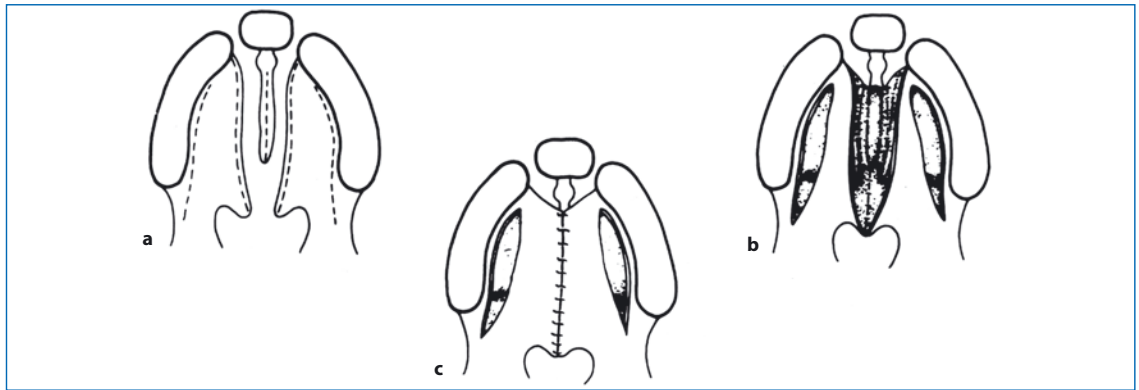
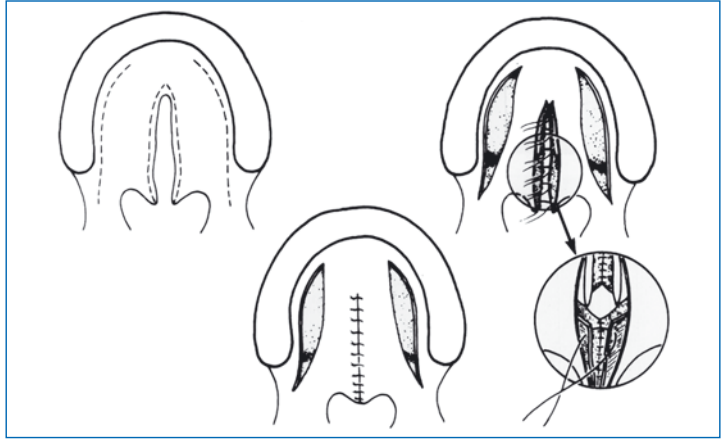
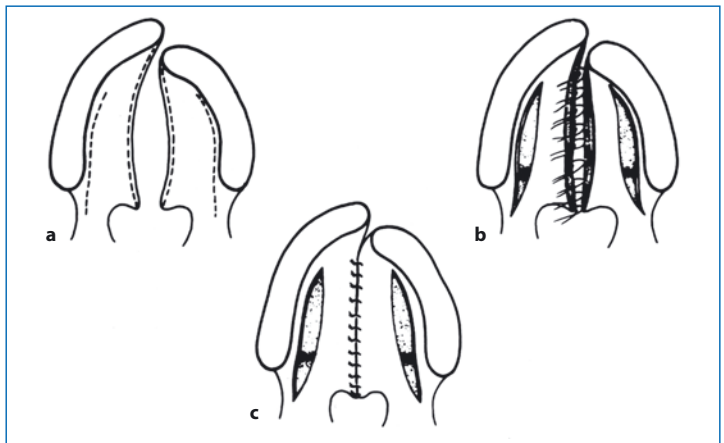


Fig. 15.10 a-c. von Langenbeck (simple closure) palatoplasty for bilateral complete cleft lip-cleft palate. The configuration of this deformity varies tremendously and line drawings can be misleading. **a** The incision lines. The inferior border of the vomer is incised and mucous membrane is elevated from both sides. **b** Mucoperiosteal flaps are elevated without anterior detachment, keeping the posterior palatine arteries intact. Extensive dissection is frequently necessary in the lateral release incision area for this anomaly when the cleft is usually wide and there is usually considerable hypoplasia. The timing of surgical closure is delayed until there is additional palatal growth at the

expense of the palatal cleft space. The nasal mucosa is closed. Two suture lines are necessary anteriorly because of the two vomer flaps. **c** Oral mucosa closure. It is impossible to close the anterior portions of the cleft with this operation. The amount of denuded palatine bone is less, however. Anterior cleft closure is performed simultaneously with a secondary alveolar bone graft. Depending on the occlusion and extent of the anterior cleft space, the premaxilla and/or the lateral palatal segments are repositioned at the time of anterior cleft closure. (Reprinted from [77] with permission from WB Saunders Co.)

Fig. 15.11 a-c. von Langenbeck (simple closure) palatoplasty for complete unilateral cleft lip-cleft palate. **a** The incision lines. **b** Mucoperiosteal flaps have been elevated, although this is not well shown in this diagram. The nasal mucosa is closed. In this anomaly, the nasal mucosa on the noncleft or medial side is continuous with the septum and vomer. More is available to manipulate. **c** Oral mucosa closure. Note that it is impossible to obtain a two-layer closure of the alveolar portion of the cleft with this operation. Lateral raw areas are relatively smaller because the mucoperiosteum is not detached anteriorly. The anterior cleft space is closed simultaneously with a secondary alveolar bone graft. (Reprinted from [77] with permission from WB Saunders Co.)



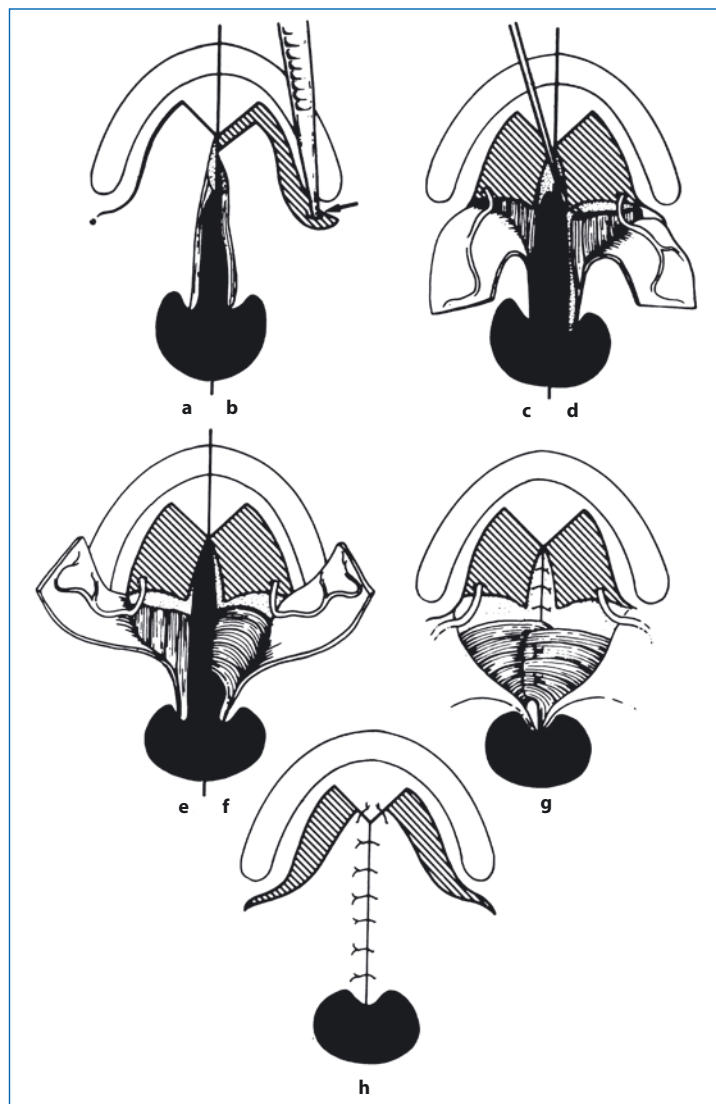


Fig. 15.12 a–h. The three flap Wardill-Kilner push-back modified after Braithwaite. **a** The margins of the soft palate cleft have been pared and the cleft of the hard palate incised along the junction of oral and nasal mucoperiosteum. The lateral incision has been made inside the alveolar ridge from opposite the canine anteriorly to a point just behind the hamulus posteriorly. An oblique incision joins the anterior end of the lateral incision to the cleft margin. **b** The mucoperiosteal flap has been elevated from the hard palate. **c** The oral mucoperiosteal flap has been turned back showing the greater palatine vessels passing from the greater palatine foramen to the flap. The mucosa has been elevated from the septum (the vomer) which is attached to the margin of the palate on this side. **d** The muscles have been detached from the back of the hard palate. The soft palate is falling away from the hard palate and becoming elongated. This is the “push back.” **e** The muscle has been freed from the overlying mucosa. **f** The muscle has been freed laterally and has been rotated medially. **g** The two muscle bundles have been overlapped and sutured under slight tension to construct the muscle sling. The nasal mucosa has been sutured. **h** Suturing of the oral layer is completed. The tips of the oral mucoperiosteal flaps are sutured to the apex of the anterior flap, indicating the degree of palatal lengthening (Reprinted [78] with permission)

Editor's Note. Many of these procedures have been followed longitudinally and have shown to be nonphysiological. Excessive scarring resulted which led to growth interference. Also, the velum has not been lengthened. Due to scar contracture the velum returned to its original length. This procedure has not

been successful in lengthening the velum but has created excessive scarring which interferes with palatal growth and development. Although the velum has been lengthened, scar contracture returns the velum to its original position

15.5 The Effect of Surgery on Maxillary Growth

Kremenak and his colleagues [24, 26, 27] reported a series of follow-up studies on beagles, based on earlier work, showing that surgical denuding of palatal bone adjacent to deciduous teeth resulted in inhibiting maxillary growth. More recent efforts by this group have focused on the contraction phase of early healing of surgical wounds on the canine palate. Olin

and associates [28] presented data based on measurements between tattoo points on wound margins on hard palates of young beagles. They noted that major interruptions in the increase of arch width coincided with the period of soft-tissue contraction. This led Olin et al. [28] to suggest that the contraction of wounds following surgery could be the first link in a causal chain leading eventually to secondary skeletal deformities.

Kremenak and associates [23] reported related evidence that the contraction seen in palatal mucosa was analogous to that reported in the healing of skin wounds. Transplanting autogenous grafts from other oral mucosa into mucoperiosteal excision wounds of the hard palate resulted in a reduction in contraction and was followed by normal or “supranormal” increases in maxillary arch width. They concluded that more research in this area was warranted.

It is becoming apparent that the factor of contractility in some types of nonmuscular connective tissue cells may be a common denominator for research on surgical wound healing, as well as for research on normal and abnormal growth and function in the craniofacial complex. There is a large body of literature dealing with advances in the understanding of contractile phenomena. Much of it is collected in a bibliographic review by Morris and Kremenak [29]. The report by Madden and associates [30] also provides an excellent overview of this area of inquiry.

Is surgery for the repair of cleft lip and palate to be determined by the age of the patient or by the palate’s morphological characteristics and the effects of surgery on the subsequent growth of the face? This was one of the driving questions being asked by surgeons in the 1940s. Slaughter and Brodie [21] and Graber [19, 20] deserve historical credit for bringing this issue to the world’s attention. Their condemnation of the results of cleft palate surgery, as mentioned, led to a new conservatism and forced a re-examination of surgical practice.

The beginning of serial facial growth studies, Graber’s [19, 20] cross-sectional study of 60 cases of mixed cleft palates of various types at various ages, had a profound effect on surgical planning. He compared jaw development of 46 operated cases with 14 cases that had had no surgery, and found that the unoperated palates resembled normal palates more than they did the operated palates. He noted that the growth of the operated palates was retarded in all three dimensions. It should be understood that it was not unusual in those days to encounter patients with a history of 35 operations. A common operation of the day involved wiring compression of the cleft segments [22]. As already noted, this procedure was notorious for its mutilating effects. Dental neglect was common, resulting in rampant caries, and the associated malocclusions were often so severe as to discourage orthodontic treatment.

Graber [19] concluded that “to minimize interference with growth centers, it seemed advisable to postpone surgical correction at least until the end of the fourth year of life, when five-sixths of the total maxillary width has been accomplished.” As a result, early surgical repair of the palate fell into disfavor, and prosthodontists came to dominate the rehabilitation of cleft palate.

Graber [19] suggested that the following questions should be asked:

1. How does the pattern of growth and development of a cleft palate individual compare with that of a noncleft person?
2. What is the effect of early and repeated surgical intervention on this pattern?
3. What happens to a tissue that has been manipulated and traumatized?
4. Does increased tension of soft tissue stimulate or depress cellular proliferation?
5. What is the growth potential of fibrous bands of scar tissue?
6. Can cicatricial (scar) bands influence the normal growth and development of the surrounding soft tissue structures and the bony skeleton?
7. How do cleft palate individuals with surgically closed clefts compare with those who have had no surgery?

Pruzansky, in 1969 at an international symposium on cleft palate held at Northwestern University Dental School, recognized the important contributions of Graber [19, 20] and Slaughter and Brodie [21] for their condemnation of the deleterious effects of traumatic surgery on midfacial growth [31] and recommended that surgery on the palate should be delayed until 5 or 6 years of age to minimize facial growth malformations. In questioning the validity of their conclusion on the timing of surgery, Pruzansky goes one step further, citing findings from his longitudinal palatal and facial growth studies [31].

I saw patients whose palates had been repaired early and yet their faces developed well and their speech was free of the stigma associated with the stereotype. On the other hand, some children, for whom palatal surgery had been delayed, did not do well at all. Their midface did not develop normally and their speech was hypernasal and unintelligible. I saw all kinds of permutations and combinations.

He went on to explain that the faultless error was in not recognizing the heterogeneity that exists within a single cleft type. Pruzansky [32, 33, 34] stressed that morphological and physiological variants in the individual child should provide the rational basis for therapeutic design, not age alone.

In the early 1920s the cleft space was considered a “hole” that needed immediate closing either by orthopedics and surgery. Early surgical closure in the first 6 months led to disastrous results. As a reaction, the timing pendulum was to swing to the opposite extreme to favor surgical closure at 5–9 years of age, when the hard palate’s growth was 90% completed. To satisfy the speech-language pathologists, obturators were worn until the cleft was closed. Based on theoret-

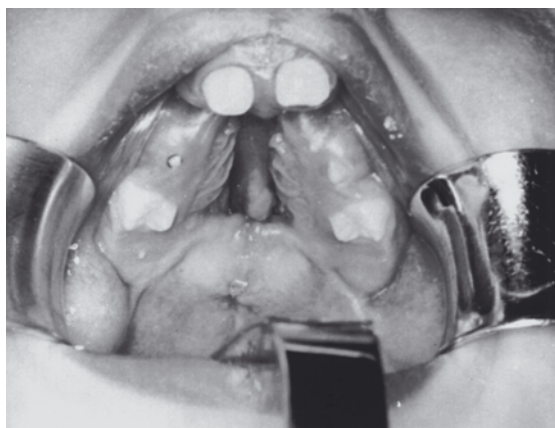


Fig. 15.13. Palatal view showing the lip and soft palate closure as a primary procedure. The hard palatal cleft is closed at a later date, usually between 18 to 30 months according to the size of the cleft space

ical and anecdotal evidence, many surgeons chose 18–24 months for closure without the use of obturators as a compromise between speech and growth requirements.

Many variations in surgical procedures arose. Gillies and Fry [35] and Slaughter and Pruzansky [36] believed it was important to delay hard palate closure to 2 or 3 years of age and chose to close the lip and soft palate first to aid speech development (Fig. 15.13). Others (e.g., [37, 38]) delayed midpalatal closure to the deciduous dentition, and Schweckendiek [39] went to the extreme and delayed closure into adolescence, favoring the need to maximize palatal growth above all else. The Slaughter and Pruzansky [36] method was associated with good speech development; the Schweckendiek delayed palatal closure procedure, and although producing satisfactory midfacial growth, left the patients with very poor speech.

15.6 Speech Considerations

Some surgeons believe that good speech requires palatal cleft closure between 6 and 9 months, the child's stage of phonemic development of articulation, and are willing to accept the trade-off of creating some maxillary growth inhibition [40]. Other surgeons believe that good speech development is not related to the age of cleft closure but depends on the growth integrity of the entire midface: the palatal vault space, the size and shape of the pharyngeal space, and the size and neuromuscular action of the soft palate and pharyngeal muscles, along with hearing and the patient's phenotype [41].

Blocksma et al. [42], Lindsay [43], Kaplan et al. [44], Krause et al. [45], and Musgrave et al. [46] found good speech with the von Langenbeck procedure in most cases. This repair gave slightly better speech results than the lengthening procedures in soft palate clefts, but the V-Y push back was superior in the more extensive clefts. However, the authors do not discuss the effects of the procedures on palatal growth and dental occlusion. Dryer and Trier [47] compared the speech results when three different palatal surgeries were performed: (1) V-Y or island pushback, (2) von Langenbeck method utilizing two bipediced mucoperiosteal flaps advanced to the midline, and (3) von Langenbeck procedure with the addition of levator reconstruction (intravelar veloplasty). The speech results after palatoplasty revealed no significant difference between children with simple von Langenbeck closure and those undergoing palatal lengthening procedures. Children with levator reconstruction demonstrated superior speech results.

As to maxillofacial development, Jolleys [48] found better speech results in his patients who had surgery prior to age 2 and found no difference in maxillofacial development. Koberg and Koblin [49] suggested that 2–3 years old is the best age for surgery without damaging maxillary development. Friede et al. [50] and Berkowitz's [51] facial and palatal growth studies do not support Ross's [52] conclusion that early (before 18 months) palatal repair provides better facial growth than does delayed hard palate repair. The opposite appears to be true because deformed palates are more closely related to early (within the first year) surgery than delayed (4–9 years of age) surgery. For example, Graber [19] reported:

In various studies of facial growth and development, it is seen that the lateral width of the maxilla is accomplished quite early in life, but the downward and forward growth is not complete until the second decade of life. Any growth disturbance induced by environmental interference would be possible in the sutural sites of proliferation for a number of years, but any appreciable withholding of the palate in width would require speech interference during the first 4 years of life.

All goals of a good speech, speech occlusion, and facial aesthetics are possible. Most orthodontists and surgeons now agree that the skills of the surgeon need to be considered when evaluating all surgical procedures, but the timing of palatal closure to avoid scarring is equally important.

Attainment of normal speech, facial and palatal development, and dental occlusion is possible without compromising one objective for another. Although speech development may benefit from early palatal closure, there are instances when the cleft space is

very wide and cleft closure should be postponed to a later age to permit additional palatal growth and allow for conservative palatal surgery. Berkowitz's palatal growth studies, presented in this text, show that an increase in palatal size with the spontaneous narrowing of the cleft space can occur early, late, or not at all; and, in rare instances, the cleft may even widen. Nonphysiological surgery causes facial and palatal deformation due to the destruction of blood supply with scar formation. To avoid these consequences, timing of palatal closure should be related to the anatomical and functional assets in the individual and not determined by age alone. Berkowitz's [51] serial studies of 36 unilateral (UCLP) and 29 bilateral (BCLP) cleft lip and palate cases with good speech demonstrated that conservative palatal surgery is conducive to good speech as well as good palate and facial development. Speech appliances, in very rare instances, may be necessary as an interim device to close off the cleft space after 2 years of age.

van Demark and Morris [53] found that early surgery (before 24 months of age – before the child begins to talk) to close the cleft space was associated with better articulation skills when the children were tested at 8 years of age, but after that age, differences were less often seen. They believed that other variables, for example, velopharyngeal competence and cleft type, were better predictors of eventual articulation skills than was age at surgery.

McWilliams et al. [54] in reviewing the literature, found better speech to be associated with earlier palatal repair (before 24 months of age). However, they noted that design flaws in the studies make it difficult to interpret results. Failure to define “normal,” “perfect,” or “acceptable” speech and the omission of information about cleft type and surgical techniques are significant shortcomings in comparing results.

Most speech-language pathologists still advocate closure of the palatal cleft before 1 year of age, believing that early closure prevents development of patterns of speech that would require prolonged and difficult therapy at a later age [54]. Most of their studies and conclusions are based on the use of only two variables: timing of surgery and speech outcome, omitting other significant variables; therefore, their conclusions must be questioned. The error in relating speech adequacy to the age that a palatal cleft was closed has only confused the issue of individualizing treatment planning based on differential diagnosis of the cleft defect.

Speech studies of individuals or a small sample of subjects are often sufficient, and sometimes essential, to finding solutions to a particular problem being investigated; however, the fact that a “correlation” exists between two variables (speech proficiency and age) does not indicate or prove causation. Although the age

at which surgery is performed may be the sole relevant variable studied, sensory function, genotype, the geometrics of the original deformity, the facial growth pattern, and the surgical procedure performed are also factors that must be considered. It is difficult, and perhaps impossible, to demonstrate the effectiveness of a treatment philosophy in a clinical setting, where many variables cannot be identified, controlled, or manipulated. Therefore, conclusions drawn from such investigations should be considered with caution [51].

Some past surgical strategies also emphasized early palatal closure and velar lengthening with the goal of resolving immediate problems with cleft space and, hopefully, preventing future speech problems associated with hypernasality. In addition, velum lengthening procedures, originally advocated in the newborn period – often without evidence of incompetent air flow control – to avoid a possible second surgical procedure, were found to be ineffective even when performed at a later age [3].

Surgery can either aid in directing the natural growth into proper channels by establishing muscle balance across the cleft defect, or it can grossly interfere with the normal developmental changes by hindering growth through interference with blood supply, introducing a scar, or destruction of growth centers. In view of the wide range of individual variations of the defect, the surgical procedure must be altered to fit the particular case, in terms of both time and technique.

Determining which surgical-orthodontic procedures are best utilized for different types of clefts is the goal of all clinicians. Unfortunately, palatal and facial growth patterns are not predictable at birth, and therefore one needs to wait to see the effects of initial treatment on the developing face before deciding what next needs to be done. When decisions to act are made under conditions of uncertainty, it is always appropriate to consider not only the probability of success but also the consequences of failure.

15.7 Surgical-Orthodontic Procedures and Sequences

It is impossible to review all of the surgical-orthodontic approaches to the closure of lip and hard and soft palatal clefts with or without the use of orthopedic plates. Some surgeons will first unite the lip, followed either by closure of the hard and soft palate in one stage, or first unite the soft palate and then the hard palate at a later age. Some even favor closing the soft palate before uniting the lip. There are as many different surgical procedures as there are differences in the timing and surgical sequence to be utilized. Some surgeons favor a vomer flap with or without a muco-

periosteal flap, others with/without a pushback, still others a mucosal tissue closure without involving the periosteum. Yet each clinical report tells of both good and bad results, usually using the number of buccal and anterior crossbites in the deciduous dentition to designate whether the procedures have failed or succeeded. Unfortunately, most reports do not have final postpubertal facial/palatal records, which are more meaningful in describing the final outcome.

15.7.1 Palate Cleft Closure Controversies Revisited

Present day corrective procedures involve surgery, the type and timing of the operation depending on whether the surgeon is a von Langenbeck soft tissue descendant, with or without presurgical maxillary orthopedics, or a Brophy “steel-clamp and silver wire” bony closure man. There is a basic philosophical conflict between the two major groups. Some surgeons employ staphylorrhaphy for closure of the cleft palate, a variation of the von Langenbeck, Furlow, or Veau-Wardell Kilner V-Y pushback procedure using mucoperiosteal flaps. They may or may not use vomer flaps to line the surface of the mucoperiosteal flap. The soft tissue is repositioned over what many surgeons consider a normal bony framework (with the exception of the cleft area) that had failed for some reason to unite in the midline. Today, most surgeons are convinced that aberrant embryonal and fetal influences force the lateral halves of the maxilla apart, making the intramaxillary width excessive. In the past, it was Brophy, today it is many McNeil disciples who utilize presurgical maxillary orthopedics, believing that the first step in habilitation is to restore what they believe to be a normal segmental relationship at a very early age. Each school of thought has intense convictions that there is only one correct approach – its own.

Four surgical approaches are commonly used to close the palatal cleft.

1. Early complete palate repair (3–9 months). Rationale: to achieve maximum speech results with a possible chance of inhibiting midfacial growth and creating severe dental occlusion. This approach favors speech above facial growth and development.
2. Delayed complete palate repair (12–24 months). Rationale: speech results are nearly as good as with earlier repair and the facial growth disturbance is less.
3. Late complete palate repair (2–5 years). Rationale: to prevent facial-palatal growth inhibition, accepting the poorer speech results. Use of a palatal obturator is needed in most cases.
4. Early lip and soft palate repair (2–9 months) and delayed hard palate repair (5–9 years). Rationale: to avoid facial and dental deformity but perhaps still achieve good speech with the aid of an obturator.

15.7.2 Scarring Inhibits Palatal Growth

Surgery to the hard and soft palate with temporary disruption of the blood supply does not, by itself, cause damages to the underlying bone. Most surgeons and orthodontists believe that the principal growth inhibitor seems to be the quantity and distribution of scar tissue that is created after surgery. When evaluating the effects of surgery on maxillary growth and development, it is necessary to consider that clefts of the palate can differ greatly in size and form at the same age due to the amount of osteogenic deficiency in the hard and soft palate and lip. The great individual variation in the relationship of the size and form of the cleft space relative to the size of the palatal segments is responsible for the differences in the amount of scar tissue formed even when the same surgical procedure is performed by the same surgeon.

A review of cleft palate surgical history clearly shows that a single mode of surgery for all cases invariably resulted in severe palatal and midfacial deformities, as well as poor speech development. Unfortunately, the same poor results still occur despite the timing of surgery and the skill and experience of the plastic surgeon. This is so because of failure to define the criteria for the timing of palatal surgery and failure to agree on which surgical procedures interfere with normal growth and development of the structures involved. Poor results were understandable when there were no standardized methods for estimating success or recording the effects of surgery on speech and facial growth and development; these shortcomings no longer exist. Nonetheless, some present-day surgical reports still do not adequately describe the original deformity; thus, the efficacy of the surgical effort cannot be evaluated.

Mapes and coauthors [55], Robertson and Fish [56], and Berkowitz (unpublished data) all concluded that nontraumatic palatal surgery accelerated the growth rate of the maxilla, helping it reach more normal dimensions in the following years. Berkowitz and associates [57] demonstrated that, in the patient with complete bilateral cleft lip and palate, after conservative palatal surgery the palatal surface area doubled from birth to the age of 1 1/2 years. Also, in an isolated cleft palate of a patient with Pierre Robin Sequence, there was a 50% increase in the palatal surface area from birth to 1 year of age. The acceleration in growth tapered off after palatal surgery in both instances. Palatal growth accelerates 6–12 months postsurgery in some cases.

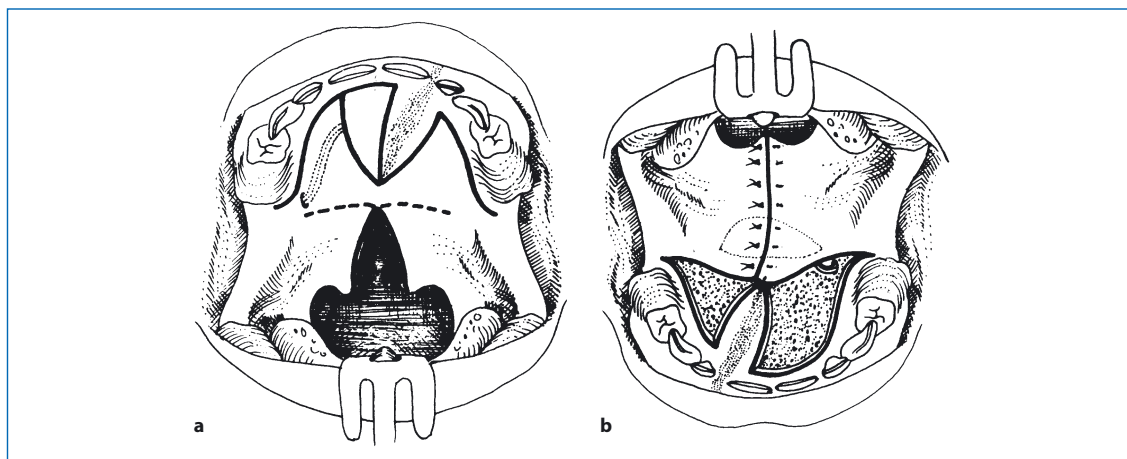


Fig. 15.14 a,b. “Island flap” for a unilateral cleft lip and palate. **a** Outline of incisions at 21 months. **b** At 21 months a V-Y pushback of the palate was achieved, leaving the V over the anterior closure untouched, and taking a unilateral island flap for nasal lining. (Courtesy of DR Millard, Jr.) Comments: The Island Flap is a variant of a V-Y pushback that is very similar to the Wardill-Kilner V-Y pushback procedure in that a large anterior area of denuded bone remains. In the “Island flap,” the anterior palatal mucoperiosteum is transposed to nasal surface creating a mucoperiosteum sandwich. This is the area that creates the transpalatal scar. Because the denuded bone heals by epithelialization becoming scar tissue, the larger the denuded area, the

more the resulting scar tissue will negatively affect maxillary growth in 3 dimensions. The maxillary deficiency usually becomes apparent in the late mixed or permanent dentition when facial appearance is affected. Lateral cephalometric studies have shown that this or other “pushback” procedures have not resulted in a net gain in soft palate length. We speculate that, in most cases, good velopharyngeal closure would have occurred even if a “pushback” had not been performed as a primary palatal cleft closure procedure. The singular lesson to learn from these longitudinal facial-palatal growth studies is the need to avoid creating large areas of denuded bone when performing palatal surgery

Berkowitz found that cases with relatively small cleft spaces prior to using modified von Langenbeck surgery grew the best after surgery. This can be interpreted to mean that the smaller the laterally placed areas of denuded bone, the less scar tissue will result with a better chance of obtaining “catch-up growth.”

Bardach [58] questions the validity of claiming that palatoplasty is detrimental to maxillary growth. He believes there is no adequate substantiation of this concept from clinical or experimental studies. However, Berkowitz’s [57] clinical report on Millard’s Island Flap pushback procedure, which creates large areas of denuded bone, has conclusively shown that this procedure deforms the palate and causes major maxillo-facial growth aberrations (Figs. 15.14–15.21). However, this study does not find fault with palatoplasties that produce small areas of exposed palatal bone. The same palatoplasty can yield different long-term results because all clefts within the same cleft type are not the same. They may have different size of cleft spaces. Unfortunately, Bardach focuses only on the surgery performed and does not consider the geometric variations in the cleft deformity as critical factors in predicting the long-term outcome of surgery.

Research is presently underway in Berkowitz’s laboratory to determine why “catch-up growth” occurs in some but not all cases even when surgery is per-

formed by the same surgeon using the same surgical procedures.

In the course of a set of related experiments, Latham and Burston [59], Latham [60, 61], and Calabrese and coworkers [62] suggested that new tissue could be induced to form in the growing face by applying appropriately controlled physical stress through neonatal maxillary orthopedics. However, no known supportive objective data have ever been presented. This subject was covered in depth in Chap. 18 titled Neonatal Maxillary.

Viteporn et al. [63] utilizing longitudinal cephalo-radiographs, showed that patients with extensive cleft palate who had a pushback procedure reached maximum growth spurt later than patients with less extensive surgery and that the surgery had an inhibitory effect on midfacial growth. They concluded that, because the sample was of the same ethnic group and received surgical treatment at the same age by the same surgeon, significant differences in midface development between the two groups should be attributed to the treatment itself. Scar tissue associated with the denuded bone left after the V-Y pushback technique, they claimed, played a major role in inhibiting forward displacement of the maxilla and in distorting dentoalveolar growth.

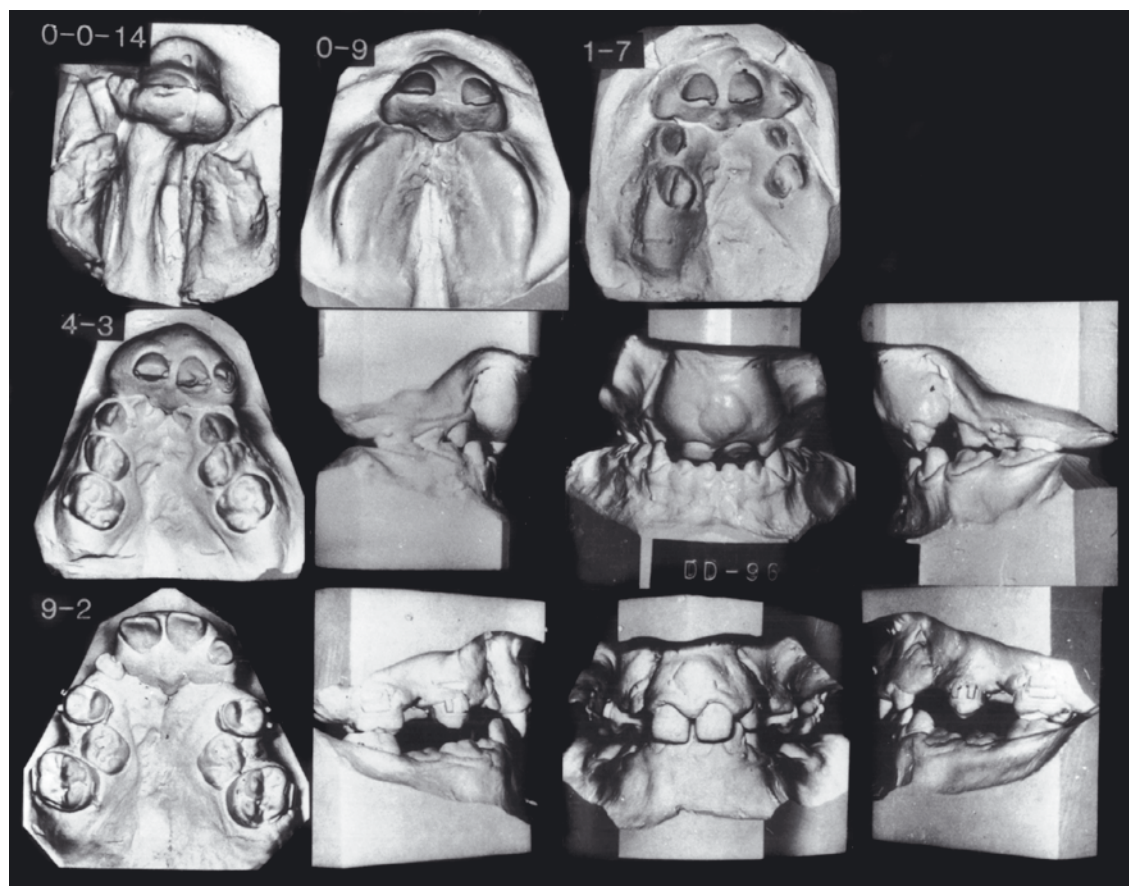


Fig. 15.15. Case No. DD-96. Serial casts of bilateral cleft lip and palate show marked palatal scarring following early “Island flap” push back. 0-0-14: At birth. 0-9: Excellent palatal arch form after the lip was united. The premaxilla-lateral palatal segments are in good relationship. 1-7: Narrow and distorted palatal arch

due to Island Flap scarring performed earlier. 4-3: Bilateral buccal crossbite with the anterior incisor are in a tip-to-tip relationship. 9-2: The palatal arch width narrowed at the line of the Island flap creating an “hour glass” shaped palate

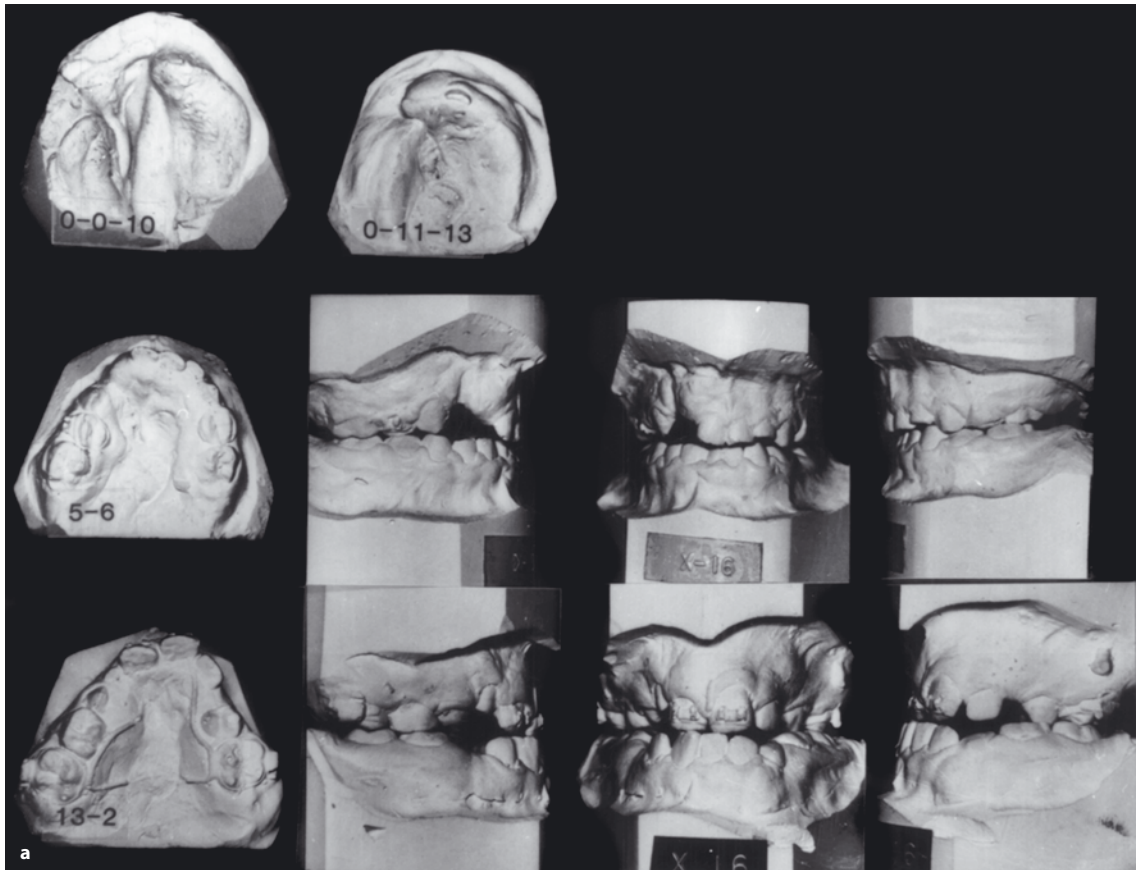


Fig. 15.16 a-d. Case JB. No. X-16. Serial palatal changes after “Island flap” leading to LeFort I maxillary advancement. **a** Serial casts. The Island flap V-Y procedure was performed at 5 months. *0-11-13*: The palate shows severe scarring and collapse. *5-6*: Even after a difficult attempt at palatal arch expansion,

the buccal and anterior occlusion are still in a tip-to-tip relationship with the opposing teeth. *13-2*: The palatal arch collapsed again due to the strong medial pull of the transpalatal scar placing the buccal and anterior teeth in crossbite



Fig. 15.16 a-d. (continued) Note that midfacial recessiveness increased as the face grew even with orthodontia and protraction mechanics and by 17-6 a Class III malocclusion existed. 17-10: After Lefort 1 maxillary advancement. 18-0: The right maxillary incisor and alveolar labial plate exfoliated as a result

of the severance of the blood supply to this area. 15-13c: A “round house” fixed bridge was utilized to replace the missing teeth and stabilize the arch form. A transpalatal removable metal strut helps to maintain the corrected arch by counteracting the medial pull of the severe scarring. Lip and palate surgery

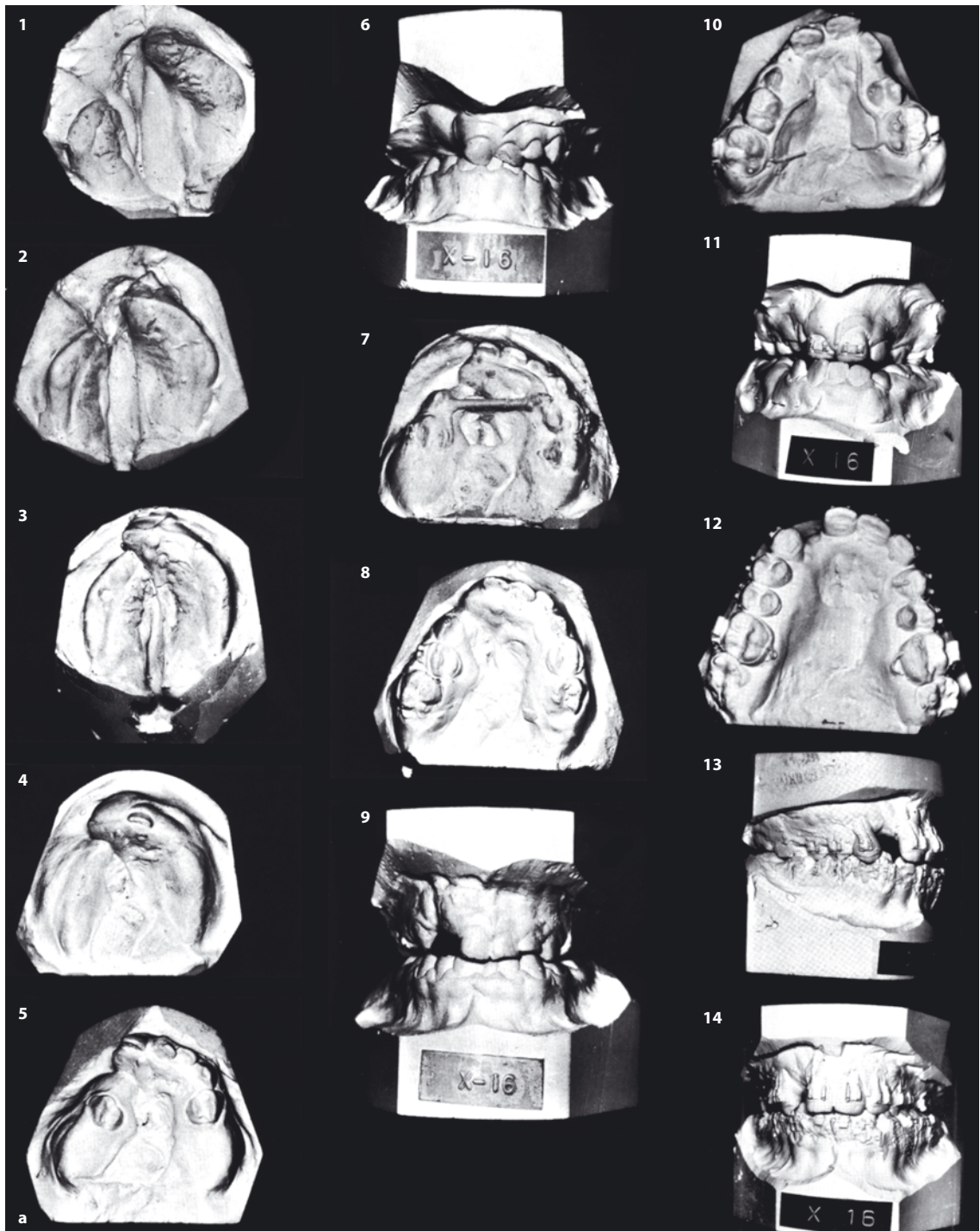


Fig. 15.16 a-d. (continued) 1 Newborn palatal cast. 2 Narrowing of the cleft space due to molding action brought on by a facial elastic worn over the lip cleft. 3 Palatal segments are in contact. 4 Further palatal narrowing after island flap pushback. 5 Severe loss of palatal vault space. 6 Right buccal crossbite. 7 Expansion appliance in place. 8 Failure to increase palatal

arch width. 9 Right buccal occlusion still tip to tip. 10 Narrowed palatal arch at 13 years 2 months of age. 11 Class III malocclusion with bilateral buccal crossbite. 12, 13, 14 (17-10) After LeFort I advancement. Excellent dental occlusion has been established. The scar tissue has been stretched, allowing for the congruency of the upper and lower arches

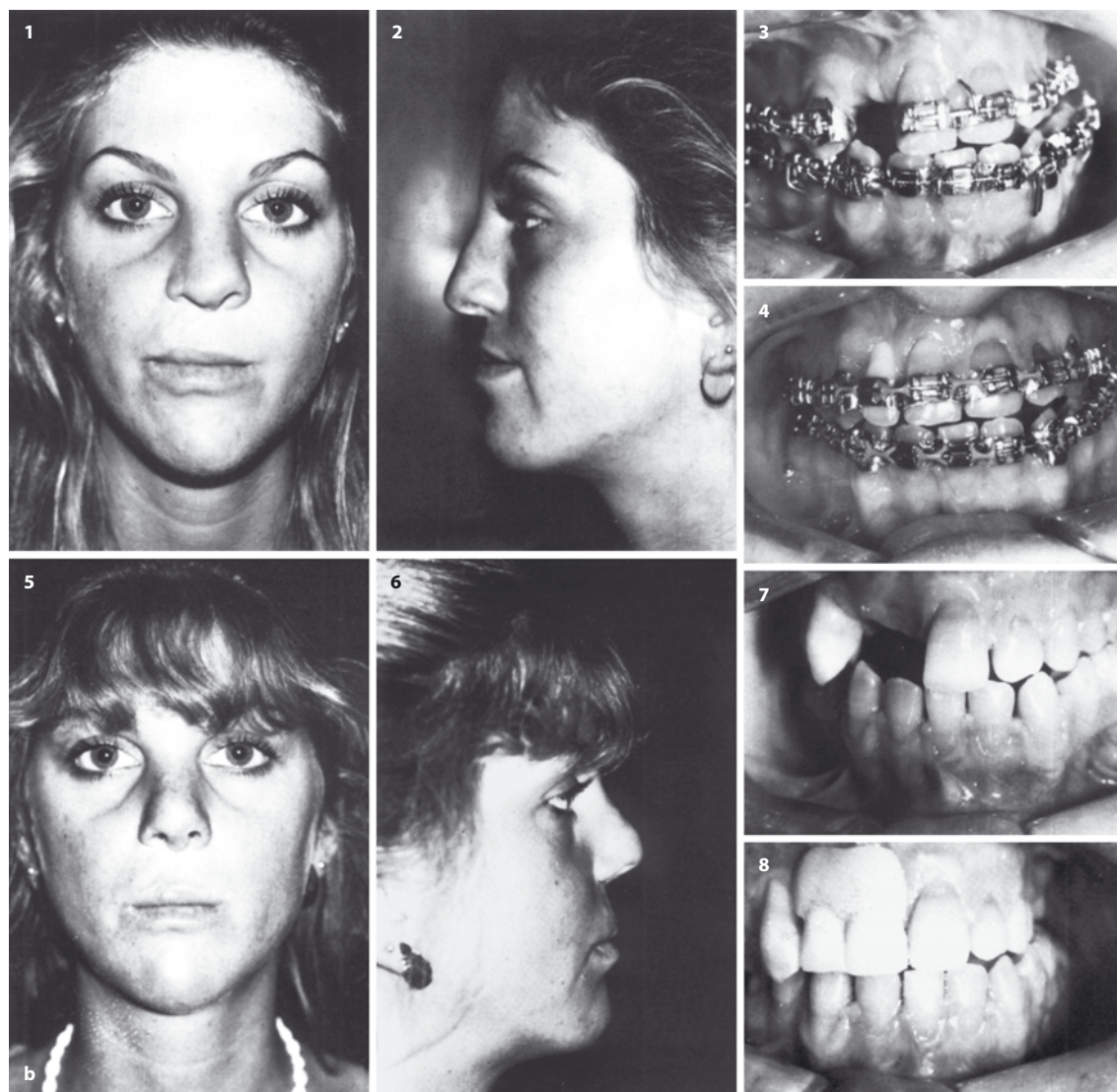


Fig. 15.16 a-d. (continued) **b** Lip and palate surgery. 1 and 2 Prior to LeFort I surgery. 3 Presurgical orthodontics. Buccal expansion and anterior advancement is unsuccessful. 4 Right lateral incisor pontic placed on the arch wire. 5 and 6 (19-3) Facial photographs after placement of a maxillary bridge and lip-nose revision. 6 Severe loss of palatal vault space. 7 (18-0)

The labile plate of bone overlying the right central incisor exfoliated because of the loss of blood supply. The posterior palatal blood supply was cut off during the soft palate pushback procedure. The central incisor had to be extracted. 8 (18-0) A temporary prosthesis replaces the missing incisors and maintains the correct arch form



J.B.

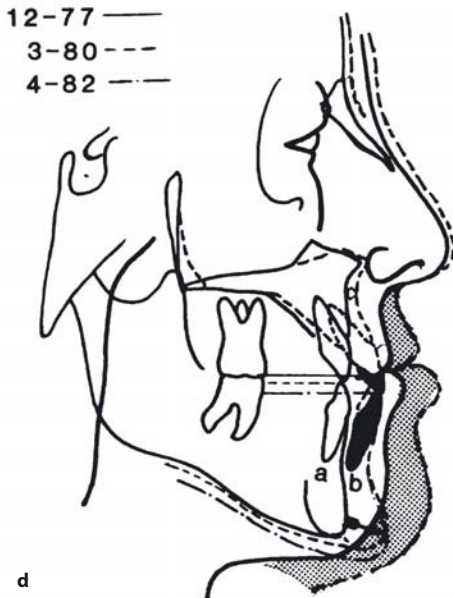


Fig. 15.16 a-d. (continued) **c** Postsurgery after LeFort I maxillary advancement to correct a retrusive midface. This followed the loss of right lateral central incisors due blood deprivation to the area. **1** Frontal and **2** lateral facial photographs. **3** Transpalatal removable cast goal arch appliance to help maintain maxillary arch size and form. **4** "Round house" bridge to maintain a functional occlusion and improve aesthetics. **d** Superimposed cephalometric tracings pre- and postmaxillary advancement show changes to the skeletal and soft tissue profile. The upper lip is more recessive than the lower lip due to the lack of maxillary basal bone coupled with additional mandibular growth. Comment: Growth disturbance caused by severe palatal scarring is three-dimensional. Although palatal osteotomies can reposition the bony segments, the force of scar contracture will prevail, causing arch collapse if it is not counteracted. This can only occur with dental bridges of various types and/or transpalatal struts [79]



Fig. 15.17. Case TO X-26. Facial changes from birth to the mixed dentition do not usually show the effect of midfacial growth retardation. The effects on facial growth becomes more evident after the postpubertal facial growth period. At birth (1), 10 months (2), 3 years (3), 6 years and 9 months (4 and 5). Frontal and lateral views at 17 years (6)

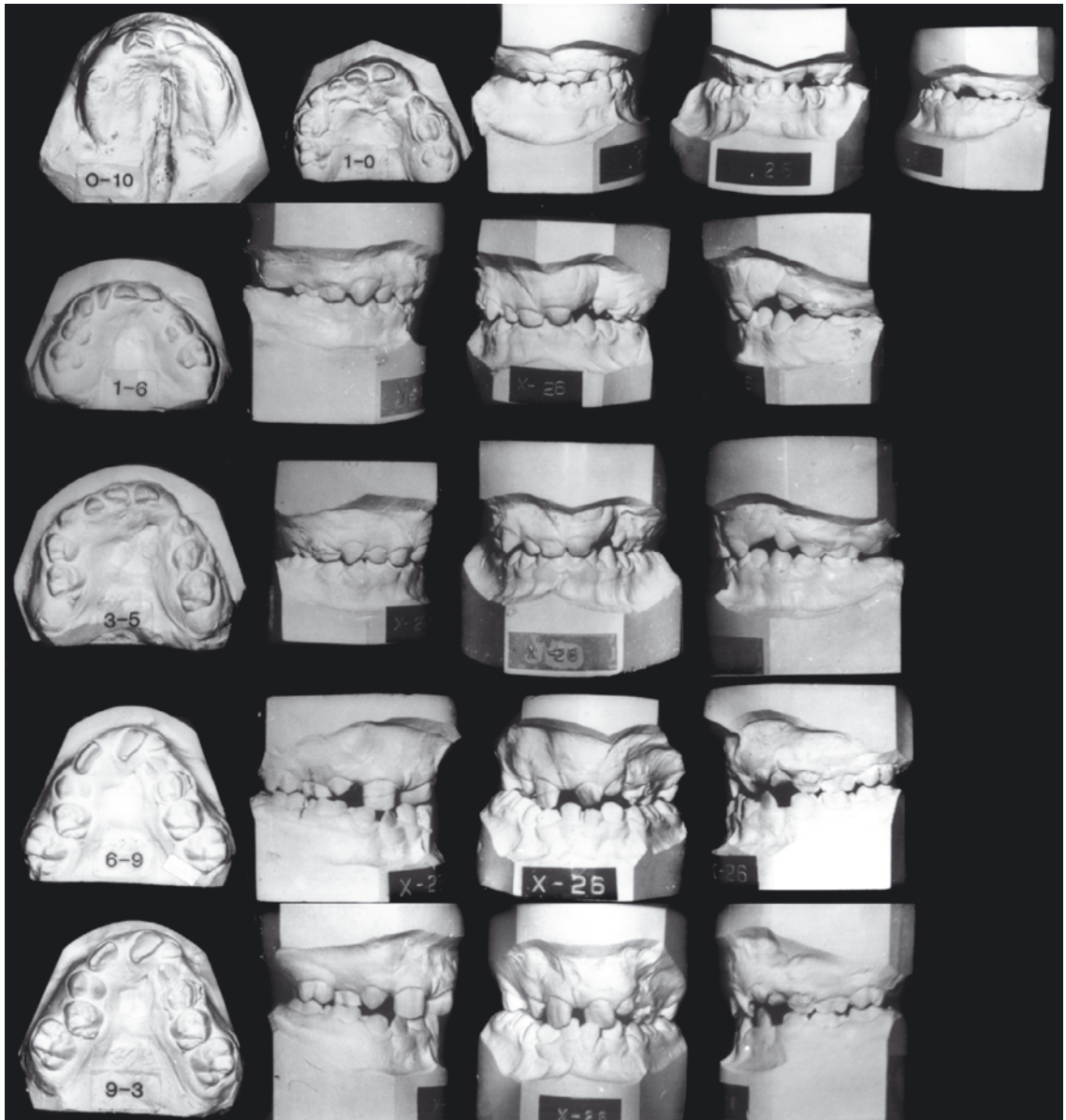


Fig. 15.18. Demonstrates the deleterious effects of excessive scarring associated with an Island flap on palatal growth, and arch form in a CUCLP. Palatal casts. 0-10: After lip surgery which brought the alveolar segment in good approximation. A small palatal cleft and good arch form before the Island Flap. 1-0: After the Island Flap and resulting collapse of the palatal

segments. The child had to protrude the lower jaw to obtain a comfortable posterior occlusion. 1-6: Anterior and buccal occlusion. 3-5: Palatal deformation becomes more apparent leading to left buccal crossbite. 6-9: Marked palatal form changes reflecting growth retardation. Further palatal and occlusal changes between 9-3

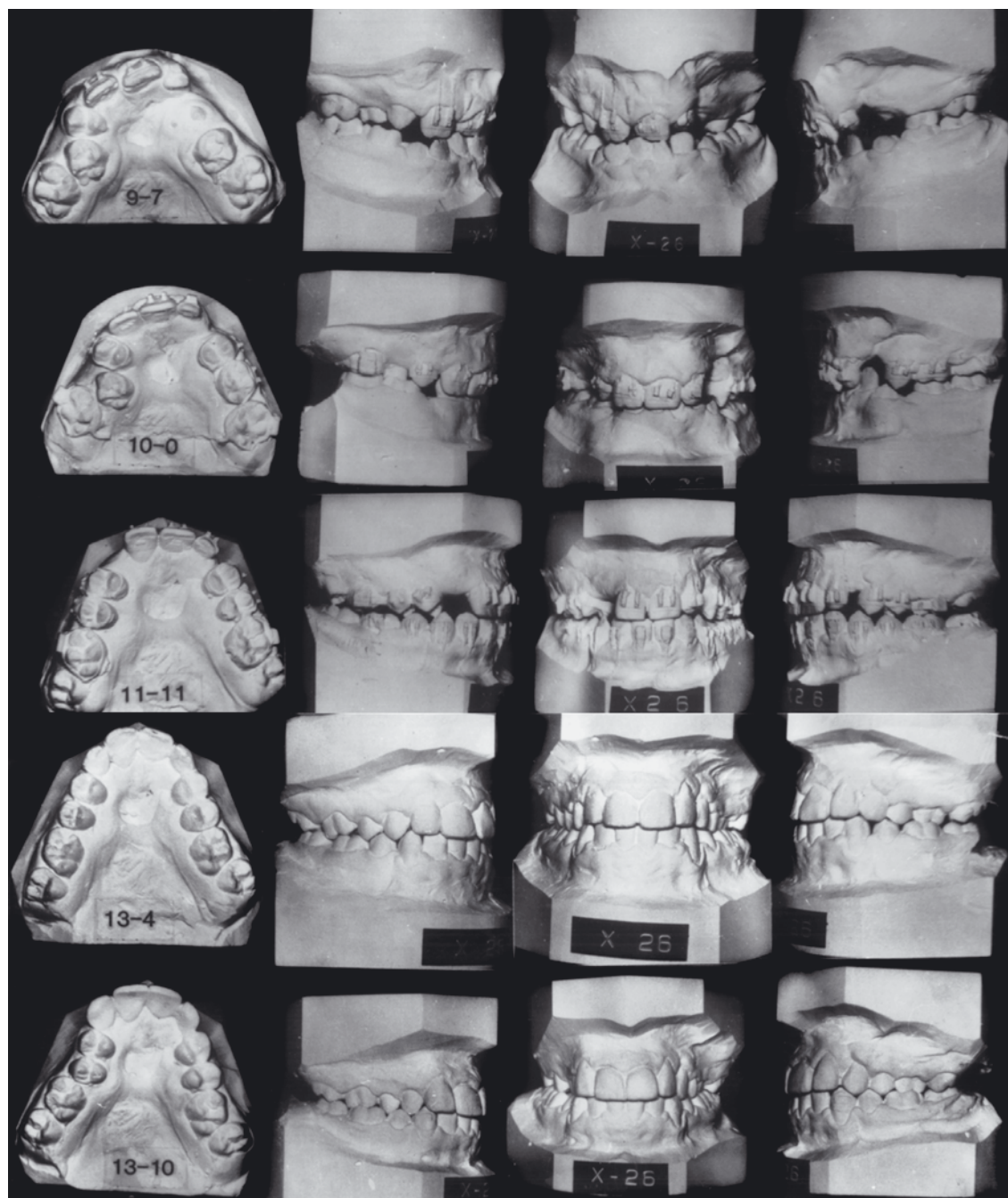


Fig. 15.18. (continued) 9-7, and 10-0 are evident. The strong transpalatal scar contracture narrows the transverse palatal width and reduces palatal growth leading to severe dental crowding. 11-11: Orthodontia was able to temporarily produce good palatal arch changes and improve the occlusion. 13-4: Both maxillary cuspids were transposed to the lateral incisor spaces. 13-10: Again the transpalatal scar tissue caused the arch form to narrow across the bicuspid and first molars. 9-7, and

10-0 are evident. The strong transpalatal scar contracture narrows the transverse palatal width and reduces palatal growth leading to severe dental crowding. 11-11: Orthodontia was able to temporarily produce good palatal arch changes and improve the occlusion. 13-4: Both maxillary cuspids were transposed to the lateral incisor spaces. 13-10: Again the transpalatal scar tissue caused the arch form to narrow across the bicuspid and first molars

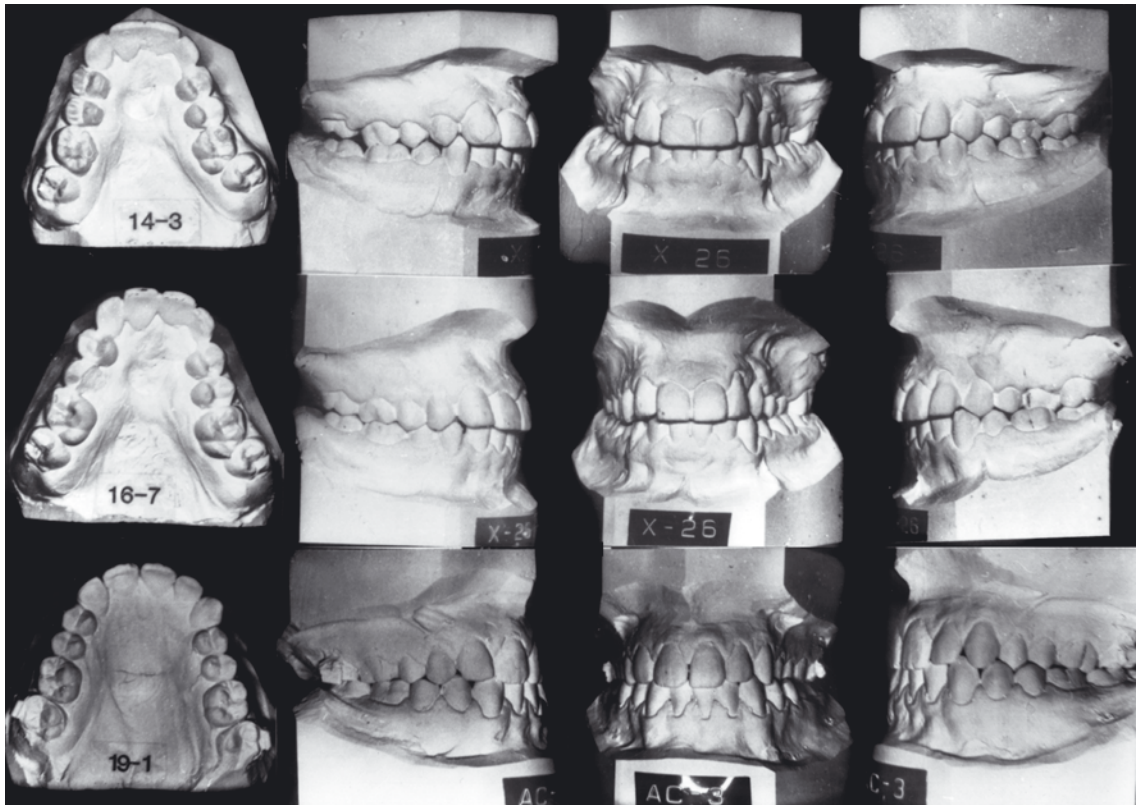


Fig. 15.18. (continued) 14-3: The narrowed palatal arch width stabilized with a good anterior overjet and overbite remaining. 16-7 to 19-1: The maxillary cuspid were positioned in the miss-

ing lateral incisor space. Millard discontinued using this palatal closure procedure as a result of the transverse scarring and the problems it created

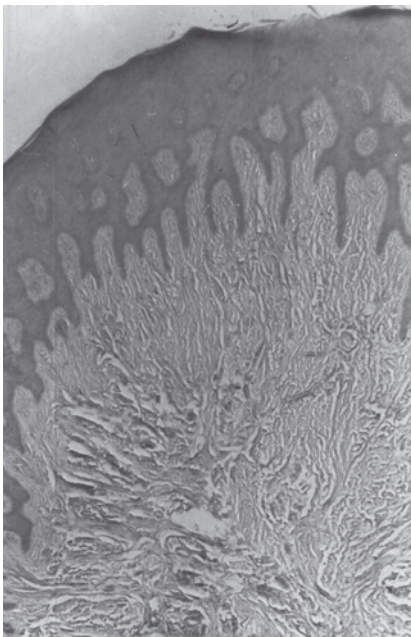


Fig. 15.19. Biopsy of transpalatal scar showing acellular band of fibrous tissue

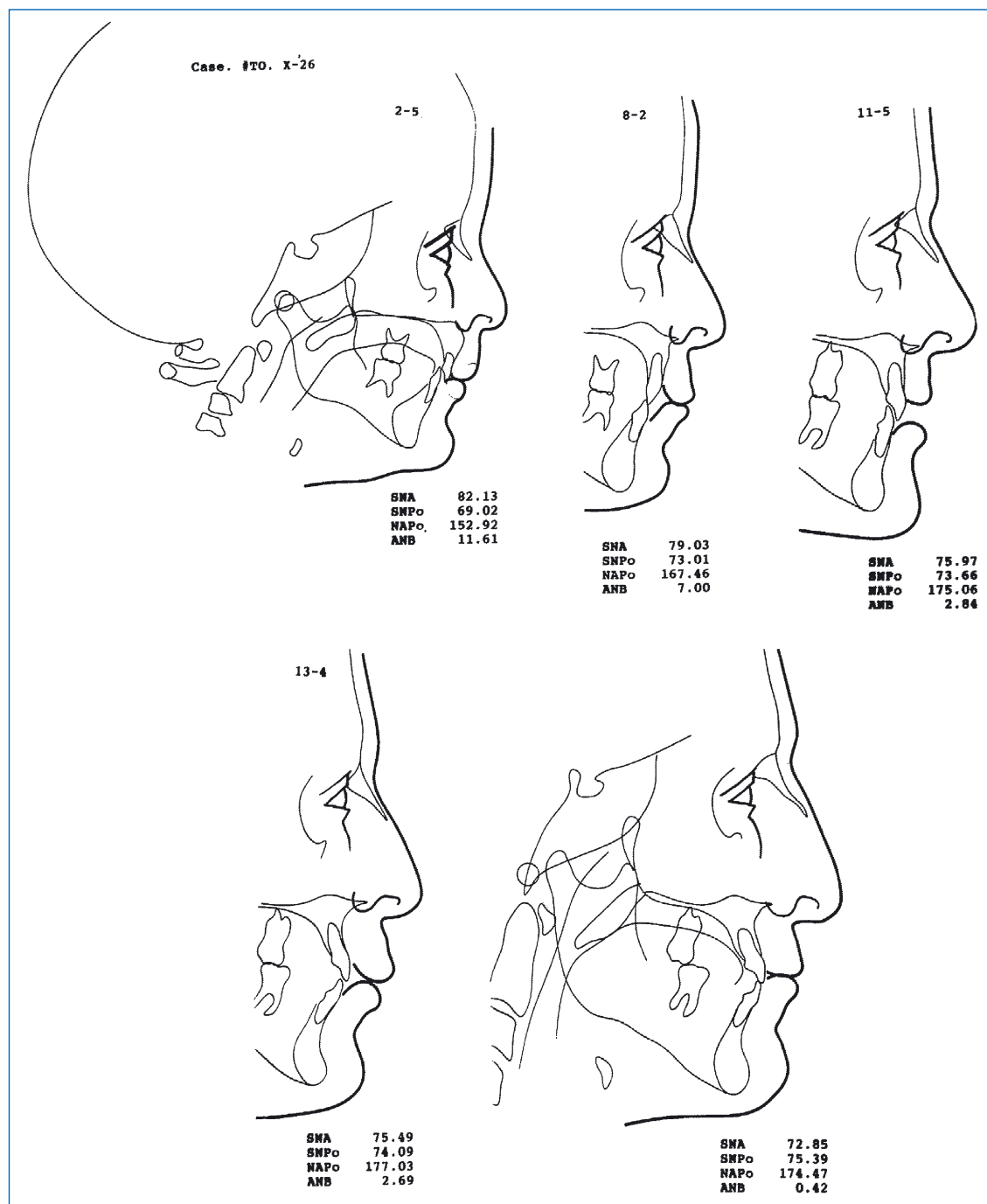


Fig. 15.20. Lateral cephalometric tracings for Case TO (No. X-26) between 2-5 and 18-0. The mandibular prominence (SNP) increased from 69° to 75° while the midfacial protrusion (SNA)

decreased from 82.13° to 72.85° during the same time period reflecting the effects of midfacial growth retardation

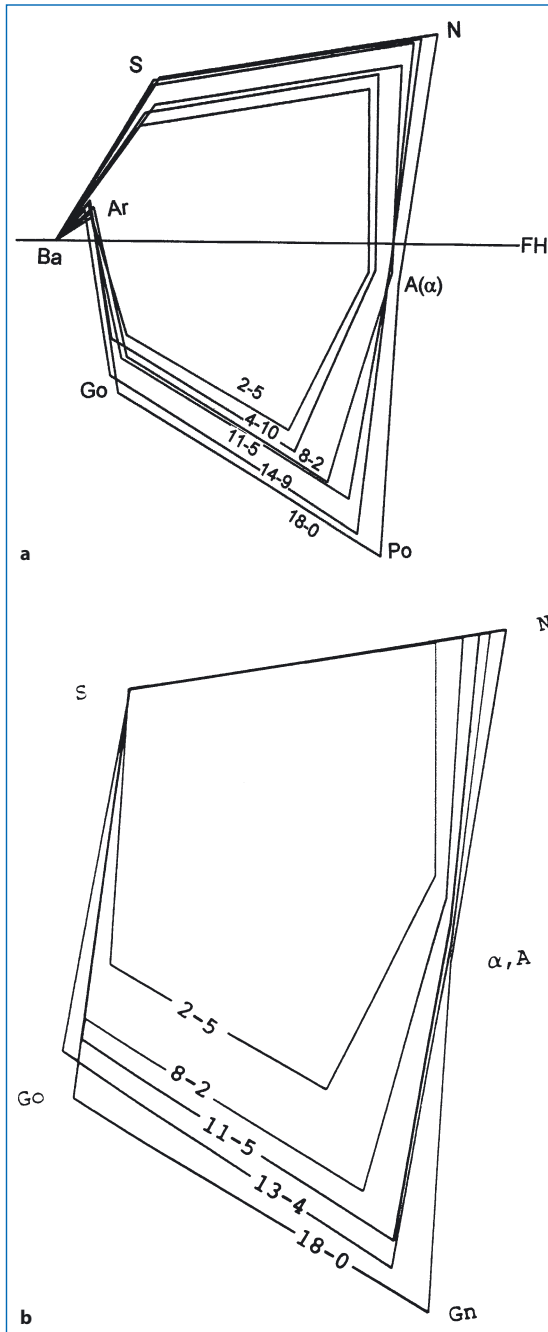


Fig. 15.21. a Superimposed polygon tracings for Case TO (No. X-26) using the Basion Horizontal method of Coben. Changes from 2-5 to 18-0 clearly show retarded midfacial growth when compared to normal growth of the upper and lower portions of the face. The soft tissue profile is able to mask the skeletal discrepancy in this instance. The upper middle and lower parts of the face are growing normally, creating a very aesthetic face with excellent occlusion. **b** Serial Cephalometric Tracings with its facial polygon. Tracings were superimposed on the anterior cranial base (SN) and registered on S. These tracings show extensive growth at the anterior cranial base and mandible. There was some midfacial growth up to 8 years but very little. Therefore, all goals of good facial aesthetics, occlusion, and speech were achieved

15.8 Dental Occlusion Associated with Early Palatoplasty Using a Vomer Flap

Dahl et al. [64] reported on the prevalence of malocclusion as seen in early mixed dentition with complete unilateral clefts of the lip and palate. The frequency of posterior lingual crossbite they found was extremely high. This might be partly an effect of the two-layer palato-vomerplasty used for closing of the cleft in the hard palate at 12 months of age. It has been shown that bone formation in the palatal cleft is common subsequent to this procedure [65]. It was supposed that the newly formed bone might act as a bony ankylosis, inhibiting the transverse growth of the maxilla. A longitudinal study, by the implant method, of patients operated on by this method substantiated this supposition. To reduce the adverse effects on transverse maxillary growth, Dahl recommended the surgical procedure be changed to a one-layer closure of the cleft in the hard palate by a vomer flap. There has not been a follow-up study to evaluate the effect of changing the procedure – whether it is the timing or the surgery that needs changing. Dahl's Danish group of treated patients were characterized by unilateral lingual crossbite, high frequencies of midline deviation, mesial molar occlusion, and mandibular overjet. There were very few cases with an anterior open bite. A tendency was seen for a difference in the sagittal molar relationship on the cleft side, as compared with the noncleft side, with distal molar occlusion being more frequent on the cleft side. This may be explained by a primary difference in the sagittal position of the two segments of the maxilla, and also as a result of secondary changes, such as lateral shifting of the mandible caused by lingual crossbite on the cleft side, medial rotation of the cleft segment subsequent to surgery, and tipping the teeth toward the cleft on the affected side.

Berkowitz's [51] serial occlusal study of complete unilateral clefts of the lip and palate, which involved the use of modified von Langenbeck procedure with a vomer flap between 18 and 24 months of age, lists an absence of distal molar occlusion with a minimal number of anterior crossbites and only a few complete buccal crossbites. By comparison with studies that do not use vomer flaps, one can conclude that the obvious cause for the deleterious occlusal results in the Dahl et al. [64] study lies in the timing and not the surgical procedure utilized because Millard [3] also utilizes a similar vomer flap with success. This conclusion does not mean that all cleft closure procedures need to be delayed until 18 months of age since narrow clefts in the palate can exist before 12 months of age. A narrow cleft suggests that less scar tissue will be created. The age of surgery is a primary variable in determining the effect of surgery on palatal development only when the width of the cleft space is also placed into the equation.

15.8.1 The Fourth Dimension of Time: Catch-up Growth

Substantial evidence from the many excellent facial and palatal growth studies (Figs. 15.22, 15.24 and Table 15.1) has determined that a "catch-up" potential exists in children with cleft lip and/or palate, which allows for reasonably good growth of the maxillary complex and face. Inherent factors alone, however, do not determine this growth. Clinicians are cognizant of the very strong influence – which quite often is adverse – of extraneous factors (e.g., iatrogenic and functional). These factors are complex and varied. It becomes obvious that surgical skill, the type of surgery, the timing of surgery, and even the sequence and numbers of surgical procedures all complicate the overall end result of facial growth in any given situation. It would appear that just the primary surgical closure of the lip and palate produces a myriad of sequences and variables to be dealt with, and they in turn introduce more sequences and variables.

To take into account both the inherent insult and extraneous factors, and to recognize and quantify the extent to which each contributes to the overall severity of the problem, is a much needed step in the correct rehabilitative effort. The inherent problem, such as the anomalous cleft, should of necessity emphasize rehabilitative efforts with minimal adverse effects on growth. At present there is no consensus as to how this could be achieved, a fact attested to by the wide variety of successful treatment regimens employed throughout the world. A long-term assessment of the approach to this problem has been brought about by the use of objective serial clinical records, such as se-

rial dental casts, by research consortium members working together [80].

From Berkowitz's serial palatal growth studies, he has deduced that the intrinsic growth potential of the maxillary processes and basal bone in cleft palate patients on the whole is slightly less in all three dimensions than in noncleft children. Various cleft types have different degrees of osteogenic deficiency, with bilateral clefts potentially being the most deficient at birth and remaining that way even as the palatal processes grow and develop. It has conclusively been shown that surgical trauma with developing scar tissue at an early age can further inhibit palatal bone growth and its forward translation within the face [66].

The structure and function of the physiological elements involved in cleft lip and/or palate, as previously explained, are not static. They exist not only within a framework of three-dimensional space, but also in a functional continuum of time. This fourth dimension of time involves the modifications induced by the processes of growth and development, which determine the ultimate nature of the congenital defect. To understand time-space relationships as they affect the individual cleft palate patient requires a longitudinal study of the patient by means of casts, photographs, and cephalometric roentgenograms.

Catch-up growth has been defined as growth with a velocity above the statistical limits of normality for age during a defined period of time. Such an increase in the rate of growth, before and after palatal surgery, with or without neonatal maxillary orthopedics, may allow the palate to attain greater size, but it may still be smaller than normal adult size. In the latter case it is called "incomplete" catch-up growth. The duration and severity of the insult (the surgical procedure used to close the cleft space in the hard palate) may positively or negatively affect the ability of the palate to recover and undergo catch-up growth.

15.8.2 The Need for Differential Diagnosis

Diagnosis and surgical treatment planning in both medicine and dentistry are most frequently dependent on the patient's age, and nature and extent of the tissue's defect. In cleft lip and palate, the timing for surgically closing a cleft palate has been traditionally based solely on the age of the patient and the onset of speech (usually between 6 to 8 months) irrespective of the physical assets and defects of the affected tissue, i.e., the relative size of the palatal cleft defect to that of the surrounding palatal tissue.

The inability to develop quantitative diagnostic criteria to facilitate a differential diagnosis for proper treatment planning is partially due to cleft palate

research programs having an insufficient number of investigative data obtained from serial maxillary and mandibular dental casts of patients starting at birth and extending into adolescence, as well as a proper instrument to measure the palatal cast's surface. Berkowitz [80] formed a consortium of four European and three American cleft palate centers to determine the relationship of the size of the cleft defect to palatal size at the time of surgery of patients that grew well into adolescence yet they all used different surgical protocols. The premise of this investigation is that data extrapolated from serial cast records will establish the relationship of the cleft defect to the palatal size and shape under the influence of corrective surgery.

Clinicians who do not have serial cast records have failed to appreciate the importance of cleft size/shape variations that exist within each cleft type at various ages, which may be crucial for making the proper decision as to when to surgically close the cleft space to avoid growth-inhibiting scarring.

15.8.3 Timing of Palatal Closure Based on the Ratio of the Palatal Cleft to the Palatal Size

A multicenter serial 3D study of the palatal mucoperiosteum medial to the alveolar ridge analysis was made of complete unilateral cleft lip and palate (CUCLP) and complete bilateral cleft lip and palate (CBCLP) casts' growth (size and velocity) changes from birth through adolescence of cases that demonstrated good occlusion, facial aesthetics, and speech. Only ones showing good outcomes were selected in order to determine which surgical protocols (timing and type) collectively produced these outcomes and what was the relationship of the size of the cleft defect to the size of the palatal segments medial to the alveolar ridges at the time of surgery. Poor outcome cases were not included because they could have been due to the surgery performed and not related to the relative size of the cleft to the palate medial to the alveolar ridge. This study was to focus on the two variables, the size of the cleft space and the size of the palate that supplies the mucoperiosteum for surgical closure. The source and number of centers in the retrospective study were; The cleft palate clinic at the University of Miami School of Medicine (CBCLP, CUCLP); University of Illinois (CBCLP, CUCLP); Nijmegen (CBCLP, CUCLP); Goteborg-Delayed series (CBCLP, CUCLP); Goteborg-Vomer series: (CBCLP, CUCLP); and Northwestern University (CBCLP, CUCLP). The Amsterdam cases were followed from birth for 48 months to eliminate the effects due to palatal surgery. This study focused on the influence of presurgical orthopedics

(PSO) at Amsterdam (CBCLP, CUCLP), and Rotterdam (CBCLP, CUCLP) which were treated with the Hotz PSOT protocol.

An exception to the well-growing cases in the Goteborg series is the Goteborg-Vomer flap cases which were included because the palate and faces developed very poorly. The purpose of including this series to compare the influence of vomer flap surgery on the palatal growth rate and size (2 mm) and to determine if palatal maldevelopment (size and velocity) values are significantly different from well-growing palates.

The control series consisted of mixed gender clefts of the lip and alveolus and/or soft palate with no palatal clefts. The growth data was the standard to which all the cleft palatal casts series were compared.

Serial palatal casts were surveyed using a highly accurate electromechanical digitizer to extrapolate surface data in millimeters, and to be analyzed using special CadCam software (CadKey). The morphometric palatal growth (size and rate of change) were compared.

Results: Berkowitz [51] concluded that:

1. The Goteborg Delayed, Miami and other institutions CBCLP and CUCLP palatal growth rates and size are statistically similar. The various clinic's cases growth (size) were less than that of the control series.
2. The 3D surface area data identified quantitative palatal parameters before palatal cleft closure that can be used as criteria for establishing a scientific basis for determining when to close the palatal cleft space.
3. Increased palatal scarring, as seen in the Goteborg-Vomer series, diminishes palatal growth.
4. Delaying all cleft closure surgery until 5+ years of age is unnecessary to maximize palatal growth.
5. Based on excellent palatal growth outcomes, the best time to close the palatal cleft space is when the palatal cleft size is 10% or less than the total palatal surface area bounded laterally by the alveolar ridges.
6. The 10% ratio generally occurs between 18 and 24 months but can occur earlier or later in some instances.
7. There is more than one good type of palatal cleft closure surgery [80].

No studies have conclusively shown that palatal growth can be accelerated by orthopedic appliances as Weil [67] claims or that "physiological" surgery with the establishment of functional occlusion will accelerate bone growth. Yet, Berkowitz [51], Krogman and associates [68], and Cooper et al. [69] suspect that catch-up growth must happen in some clefts after surgery due to the amount of additional growth that occurs, as

measured by changes in palatal surface area (the area between the alveolar ridges and limited posteriorly by the end of the hard palate).

Berkowitz's palatal growth charts have shown a marked increase in palatal surface area in the first 2 years before palatal surgery. After palatal cleft closure between 18 and 24 months, there appears to be a growth hiatus, creating a plateau in the growth graph; however, 6–12 months postsurgery there is frequently a second period of growth acceleration which extends to 6 years of age. The palatal surface area continues to increase as the molars develop and erupt into the arch.

Berkowitz and Millard palatal cleft closure timing is based on the size of the cleft defect not fixated on the patient's age alone. Some surgeons have found a “middle of the road” treatment plan to be around 12–18 months of age and discount the size of the cleft space.

15.9 Good Speech Is Dependent on a Palate of Relatively Normal Size and Shape

Sally Peterson-Falzone et al. [70] have written that malocclusion needs to be considered during the early speech learning years. She points out that the dental and orthodontic literature contains fairly consistent information regarding the effects of dental problems and malocclusions on speech.

A Need For Differential Diagnosis and Treatment Planning: A careful review of cleft palate surgical history makes it clear that a single mode of surgery based on age alone for all cases frequently results in severe palatal and midfacial deformities as well as poor speech development.

In general, this literature tells us that dental and occlusal problems are more likely to be causative factors in speech problems (1) when they occur in combination rather than singly, (2) when they are present during the speech-learning years as opposed to later years, and (3) when they influence the spatial relationship between the tip of the tongue and the incisors [70]. The literature also indicates that speech problems are fairly common when there is a restriction in the size of the palatal vault, which is more apt to be found in Class III occlusions compared with Class II [70]. Children with clefts are obviously vulnerable to restriction in size of the palatal vault and the possibility of Class III occlusions due to the presence of dental or occlusal problems (possibly several at one time) during the speech learning years. The question is: Will the speech problems diminish as the dentition or occlusion improves?

This statement convincingly acknowledges that good speech development is contingent on good tongue–teeth relationships within a normal vault space of proper volume.

Early palatal surgery (before 1 year of age in most instances) may not always jeopardize palatal and facial development provided conservative surgical methods are employed when the cleft space is sufficiently small.

There are no longitudinal palatal growth studies which show that overlapping palatal segments in CB-CLP and CUCLP are incapable of growing and developing normally. The reverse is true! Many longitudinal palatal and facial growth studies show that overlapped palatal shelves have the potential of growing well when there is minimal scarring of the palatal mucosa. Why is such a big fuss being made by those who perform presurgical orthopedics about the need to prevent “collapsed” arches? The use of the word “collapse” has a strong negative connotation which infers that a problem exists which should be prevented from occurring and if it does happen it needs correction.

In fact, there are some benefits associated with the narrowing of the cleft space due to the unimpeded medial movement of palatal segments. They are: (1) The perpendicular plates of the sphenoid, which are displaced laterally in the pharyngeal space at birth, become more normally positioned after lip surgery. (2) A reduced cleft space may encourage better tongue posturing and with it improved speech development and feeding. This still needs to be proven. Should a dental crossbite result, it can be easily corrected in the deciduous dentition using simple orthopedic forces although necessitating prolonged retention. Curiously, in many cleft palate centers, prevention of maxillary constriction (collapse) is still a major focus of attention, despite the fact it has never been shown to cause long-term difficulties associated with growth inhibition.

Unfortunately, when the clinician's treatment protocol is designed to prevent “collapse,” the team's focus is diverted and less attention is given to appreciating the heterogeneity of surgical procedures employed, different standards for selection of patients, and lack of standardization of evaluation techniques. Indeed, with a change in focus to conservative treatment planning, most surgeons now believe there will be a greater chance for improved long-term outcomes.

The developmental age of the infant at the time of the insult and the nature of the insult itself (extent of denuded bone left after surgery) will affect the ability of the infant to achieve complete catch-up growth. The case in Figs. 15.22–15.25 demonstrates the catch-up growth phenomenon. Berkowitz suggests that the effects of accelerating or restricting growth are not seen



Fig. 15.22 a-r. Case CM (AC-33) illustrates excellent palatal growth in a CUCLP treated conservatively without presurgical orthopedic treatment. Lip closed with a Millard Rotation Advancement at 7 months. Soft palate united at 16 months and

the hard palatal cleft closed at 24 months of age using a modified von Langenbeck. **a** Newborn: 24 days. **b** After lip adhesion at 3 months. **c** After definitive lip surgery at 7 months. **d** 4 years, 10 months. **e** and **f** 5 years. **g** and **h** 8 years, 4 months

in all dimensions throughout the palate. He suggests that the features most likely to be affected have very strong clinical implications in deciding at what age the palatal cleft should be closed. It is expected that

morphometric analysis of the changing form and size of the palate will permit some insight into the appropriate surgical procedures (age and type) to prevent growth retardation.



Fig. 15.22 a-r. (continued) **i** and **j** Left cuspid crossbite. **k** Palatal view showing mesioangular rotation of the left palatal segment. **l** Palatal expander after expansion. **m** 17 years. **n** 18 years. **o**, **p**, **q**, and **r** 17 years. Comments: A mandibular central incisor was extracted to allow a proper overjet-overbite relationship. Even with the extraction of a mandibular incisor it was difficult to create more of an incisor overbite relationship due to the relative palatal midfacial recessiveness. Note the short midfacial vertical dimension with a relatively long lower facial height

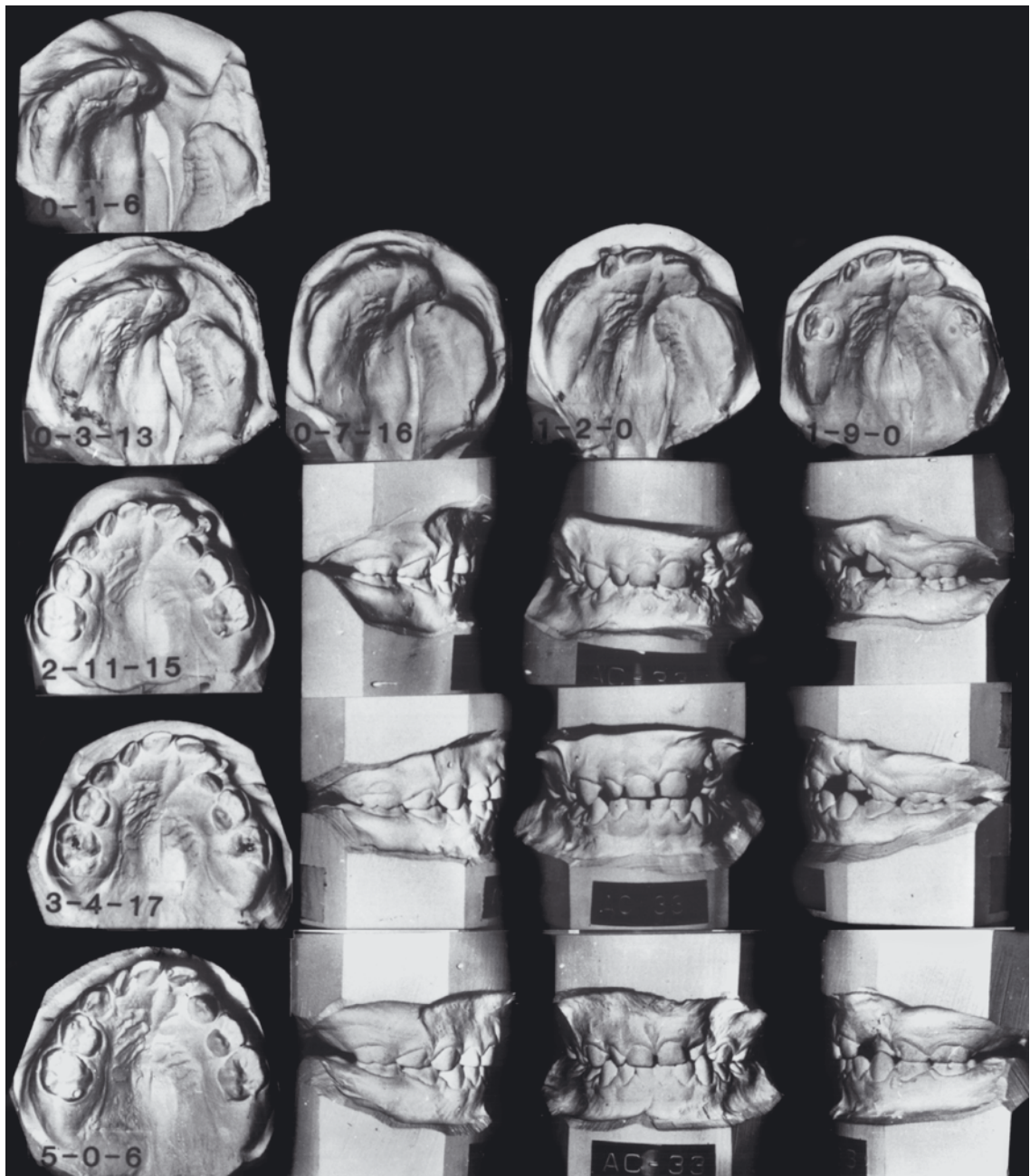


Fig. 15.23. Case CM (AC-33) serial dental casts. 0-1-6 at birth, 0-3-13, 0-7-16, 1-2-0, and 1-9-0. With lip closure the palatal segments moved together with a slight overlap of the smaller cleft

segment by the noncleft segment. 3-4-17, 5-0-6, and 5-7-0. Only the left deciduous cuspid is in crossbite

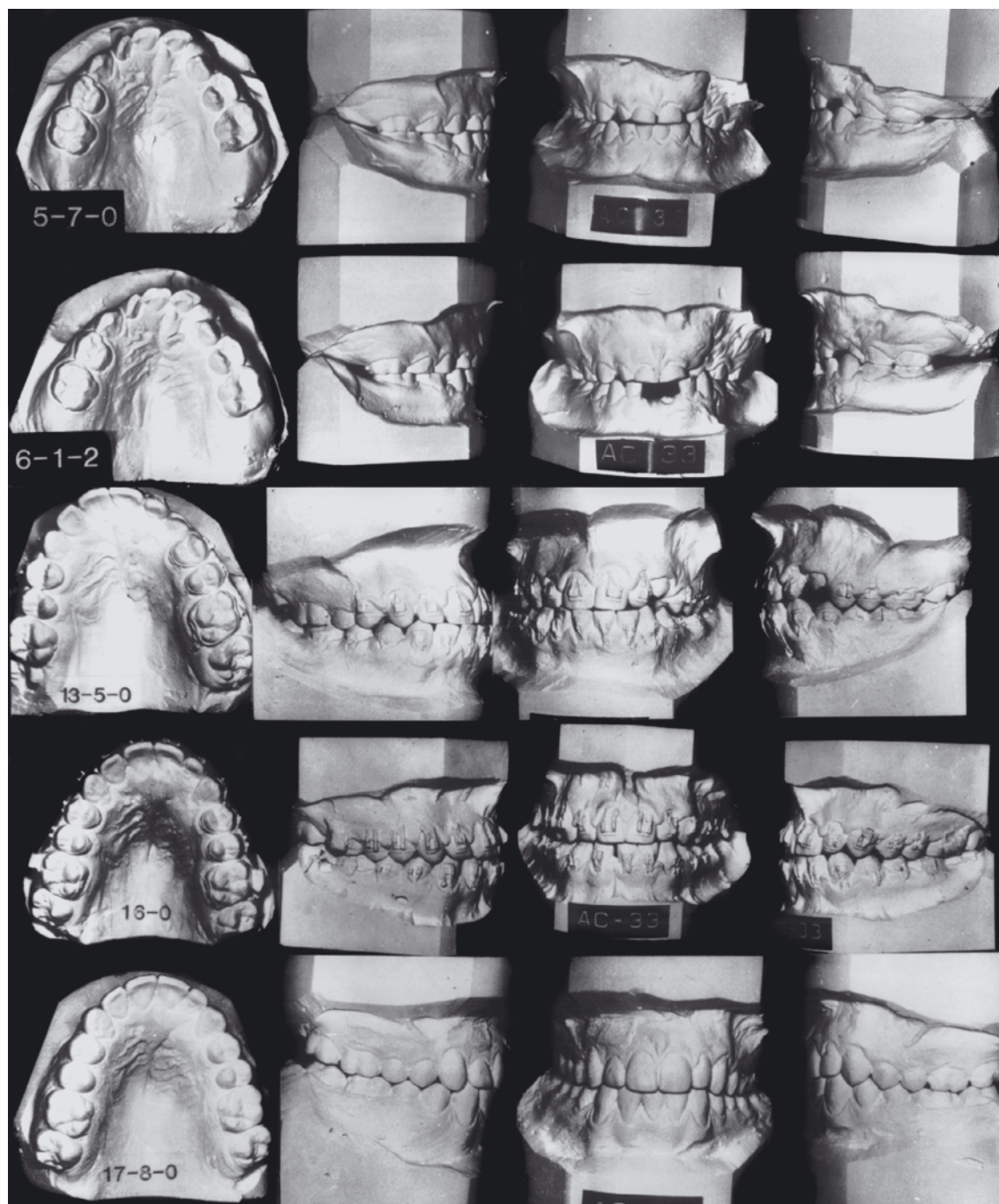
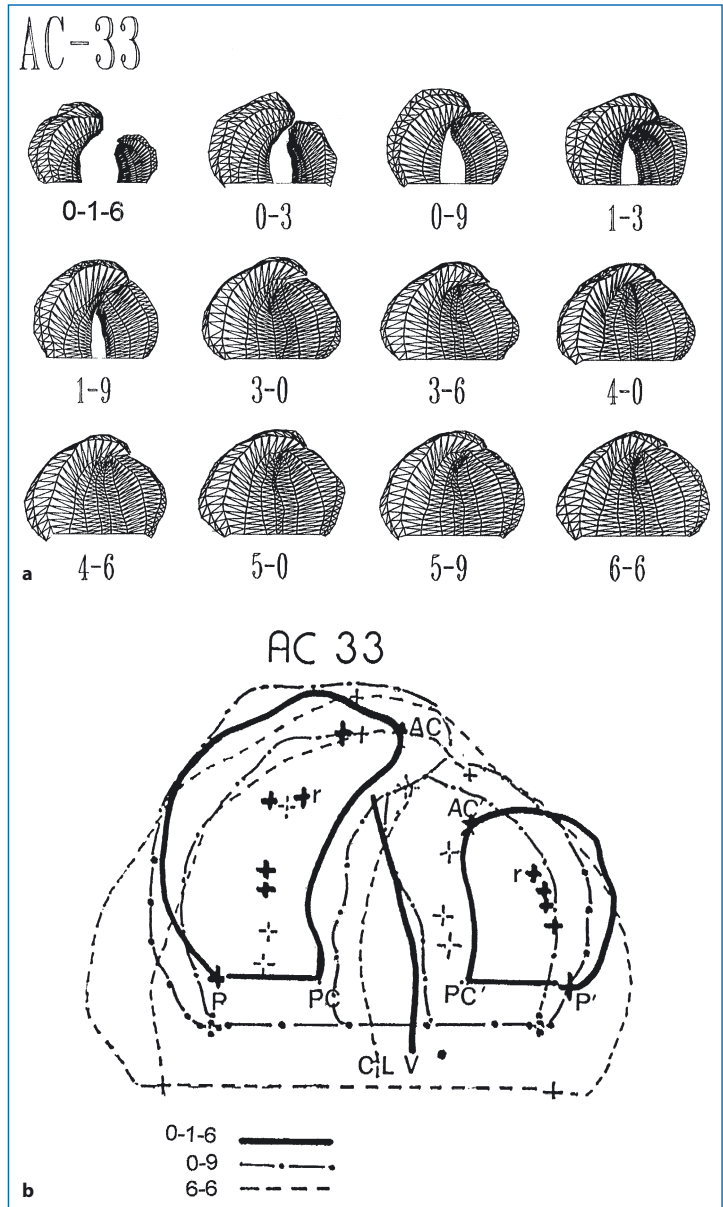


Fig. 15.23. (continued) 6-1-2. Excellent vault space to accommodate the tongue, as attested by the good dental occlusion. The deciduous cuspid crossbite did not cause a functional problem nor did it inhibit palatal growth of the lesser segment. 13-5-0. The left lateral incisor and cuspid erupted through the

secondary alveolar bone cranial bone graft performed at 8 years and 5 months of age. Tip-to-tip anterior occlusion. A lower central incisor was extracted to permit a proper overjet overbite relationship to be established. 16-0 and 17-8-0. Excellent anterior and buccal occlusion with ideal palatal vault space

Fig. 15.24 a,b. Case CM (AC-33) from 0-1-6 to 6-6. **a** Computer-generated outlines of serial casts drawn to scale. Three-dimensional serial palatal growth representation using an electromechanical digitizer and CadCam software shows the degree and location of growth changes and the relative changes in cleft space size. **b** Superimposing computer-drawn cast tracings taken at 0-1, 0-9, and 6-6 on the palatal rugae and registered on the vomer point (V) as it crosses the *P-P1* (postgingival point) line which is posterior limit of the hard palate. This graph demonstrates the great increase in palatal size and direction of palatal growth that occurs when physiological surgery is performed (see Chap. 16). *P* and *P'*: Postgingival. This landmark is comparable to point PTM (pterygomaxillary fissure) which is found between the maxillary tuberosity and the perpendicular plate of the sphenoid. *Pc* and *Pc'* – Landmark on line *PP'* at the cleft space. *Ac* and *Ac'* – Landmark at the anterior most point of the alveolar ridge of the cleft space. *V* – Point at which the vomer crosses the *PP'* line. This figure shows palatal molding changes that follow lip adhesion. Most of the growth occurs posteriorly to accommodate the developing molars



When asked at what age he operated, Pruzansky [71] replied:

We still hear the same question today. Is chronologic age the parameter that really matters, or is it morphologic age and physiologic fitness? We should be asking whether the tissues are adequate in quantity and quality. Is the geometric relationship of the malformed cleft parts to the contiguous anatomy favorable or unfavorable for reconstruction? What is the relationship of the malformed cleft parts to the contiguous anatomy

which, in turn, may be anomalous? What are the changes incident to growth? Are the parts static in their deficiency or does this deficiency diminish in time?

LaRossa [72] State of the Art in Cleft Palate Surgery, a two-layered, tension-free closure of the hard palate is advocated using a vomer flap for nasal lining and mucoperiosteal flaps that leave minimal amounts of exposed palatal bone. This approach seems to result in the least amount of growth retardation and a reduced incidence of fistula formation.

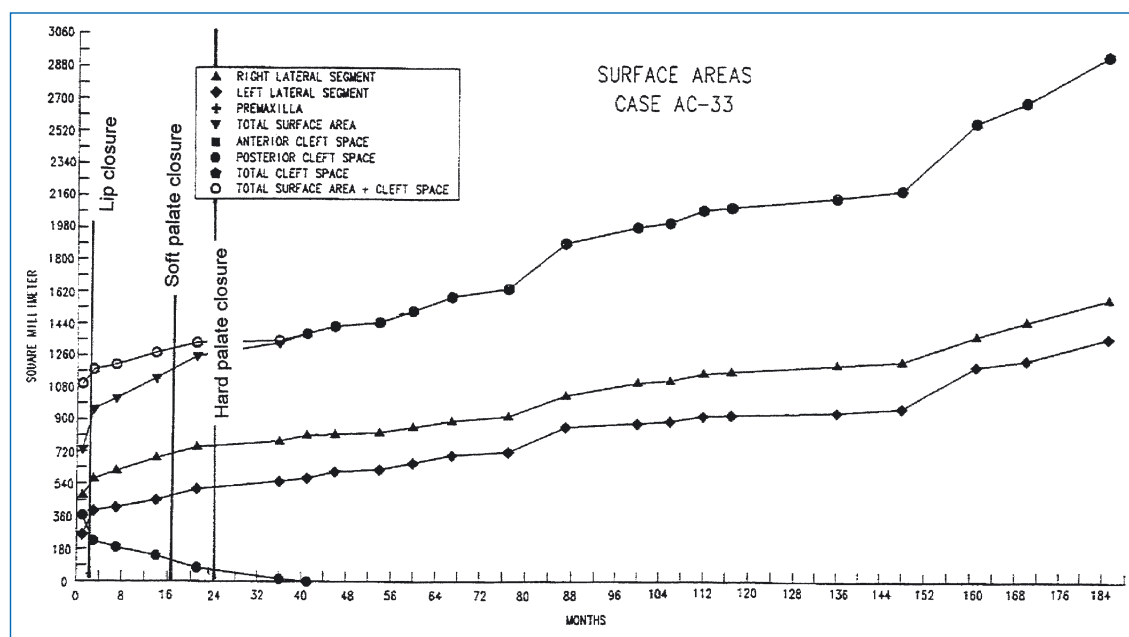


Fig. 15.25. Case CM (AC-33). The palatal growth chart shows a very rapid period of acceleration occurring the first year. During the first 8 months the palatal surface area (bordered by the alveolar crests) increased by 45%. Growth gradually slowed down after 20 months. Comment: Note that both lateral palatal segments grew at parallel growth rates. The growth acceleration

curve after palatal surgery was the same as that before surgery. The cleft palatal segment shows two growth periods; at 80 and 152 months. Since rapid palatal growth occurs in all cleft palate children during the first year, caution should be exerted when closing the cleft spaces at this age period to avoid creating excessive scar tissue when its negative effect would be greatest

Table 15.1. Case. CM (AC-33). Surface area of CUCLP from 21 days to 15 years and 5 months of age. These numbers were used to create the Growth Chart in Fig. 15.25.

Age	Skeletal Area			Cleft Space Total	
	RLS	LLS	Tot	Post	SA+CS
0-0-21	474.7	259.3	734.0	364.9	1098.9
0-3	567.6	389.6	957.2	223.1	1180.3
0-7	611.3	407.7	1019.0	187.5	1206.5
1-2	682.5	448.4	1130.9	141.7	1272.6
1-9	743.2	510.4	1253.6	75.6	1329.2
3-0	775.3	552.1	1327.4	13.9	1341.3
3-5	809.6	570.8	1380.4		1380.4
3-10	814.3	606.5	1420.8		1420.8
4-6	825.5	618.9	1444.4		1444.4
5-0	855.1	654.4	1509.5		1509.5
5-7	890.0	698.8	1588.8		1588.8
6-5	917.3	718.4	1635.7		1635.7
7-3	1033.5	857.7	1891.2		1891.2
8-4	1103.6	875.8	1979.4		1979.4
8-10	1116.6	888.7	2005.3		2005.3
9-4	1157.8	918.8	2076.6		2076.6
9-9	1168.0	924.4	2092.4		2092.4
11-4	1204.9	938.8	2143.7		2143.7
12-4	1223.3	962.6	2185.9		2185.9
13-5	1366.9	1197.5	2564.4		2564.4
14-2	1448.8	1231.7	2680.5		2680.5
15-5	1575.8	1357.7	2933.5		2933.5

Note: RLS, Right Lateral Segment; LLS, Left Lateral Segment; Tot, Total Surface Area; SA+CS, Bony Surface Area + Cleft Space Area.

15.10 Facial Changes in Successfully Treated Cases

15.10.1 Lateral Cephalometric Results from the Oslo Team

Semb's group [73,74,75] conducted a serial lateral and frontal cephalometric study of 90 cases from the Oslo archives with bilateral cleft lip and palate. Since 1962 the treatment procedure has involved uniting the lip and hard-palate cleft space closing in two stages. No presurgical orthodontics were utilized since the surgeons and Bergland, an orthodontist and past director of the program, believe that any bilateral cleft lip can be closed without presurgical palatal manipulation.

In the period spanning 1950 to 1960, a von Langenbeck procedure was performed to close the hard-palate cleft between 3 and 4 years of age; after 1960 the timing of the closure was reduced to 18 months of age. Secondary alveolar bone grafting using cancellous iliac crest bone was performed prior to the eruption of the permanent canine teeth [76]. Twenty-five percent of the updated cases needed superior-based pharyngeal flaps which were performed, if possible, before the child started school. No orthodontics were utilized in the deciduous dentition. Protraction headgear in the mixed dentition was used in one third of the cases. Fixed retention was necessary in all cases. Results showed that: (1) The maxilla progressively receded over time; (2) the mandible was retrusive with a steep mandibular plane with an increased gonial angle; (3) anterior lower-face height was elongated and posterior facial height reduced; (4) the facial growth pattern was notably different from the normal Bolton Standards: (a) male and female facial growth patterns were similar except that the males' linear dimensions were larger; (b) the prominent premaxilla would gradually realign in the preschool years; and (c) surgical premaxillary setback was never required.

The overarching thesis of this chapter favors consideration of the total emotional and physical health of the child with a cleft, based on the desired attainment of a cosmetically attractive face, and adequate dental function and respiration, as well as speech. Many surgical, medical, and dental therapies may be necessary in the best-treated cases. As long as the surgeon individualizes the treatment plan, taking care to do no harm to growing structures, all goals are obtainable.

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16 Diseases of the Ear in Children with Cleft Palate and Craniofacial Anomalies

SYLVAN E. STOOL

The high prevalence of otitis media in infants and young children with cleft palate and other craniofacial anomalies has been documented in numerous publications in the past 20 years [1–3]. Prior to that time, the high incidence of hearing loss and otorrhea had been observed in older children and adults as a major sequela of the cleft palate deformity.

The initial observations of ear disease in infants was stimulated by the development and use of the operating microscope. Further technological developments such as auditory brainstem response testing, the acoustic bridge, and endoscopic optical systems are now providing more information regarding anatomical and functional abnormalities associated with clefting. This chapter will review diagnostic methods, etiologic factors, and some aspects of treatment and outcomes.

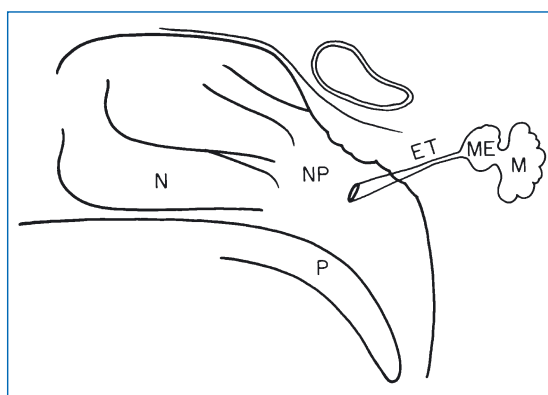


Fig. 16.1. The pneumatic system of the ear. Abnormality of the nose, nasopharynx, mastoid or Eustachian tube may affect the middle ear

16.1 The Auditory System

The ear can be divided into the conductive or pneumatic portion. The ear is intimately related to the airway and is part of a system that contains air (Fig. 16.1). Abnormalities of the nose and palate such as occur in cleft palate children may have a secondary effect on the pneumatization of this anatomical region. It is therefore important, when considering factors that may affect this pathogenesis of ear disease, to include a complete examination and evaluation of these structures. However, for purposes of this chapter, we will concentrate on the methods of diagnosis and specific findings of the middle ear.

16.1.1 Observations of the Tympanic Membrane

The diagnostic otoscope is a primary tool for the physician's evaluation, and it is important for the health-care provider who examines children with cleft palates to be familiar with the principle of pneumatic otoscopy, because middle-ear effusions are so frequent and will decrease the mobility of the tympanic membrane (Fig. 16.2). The external canal must be free of debris, because many cleft palate children have cranial and facial abnormalities associated with small external canals, and small amounts of cerumen may obscure the tympanic membrane.

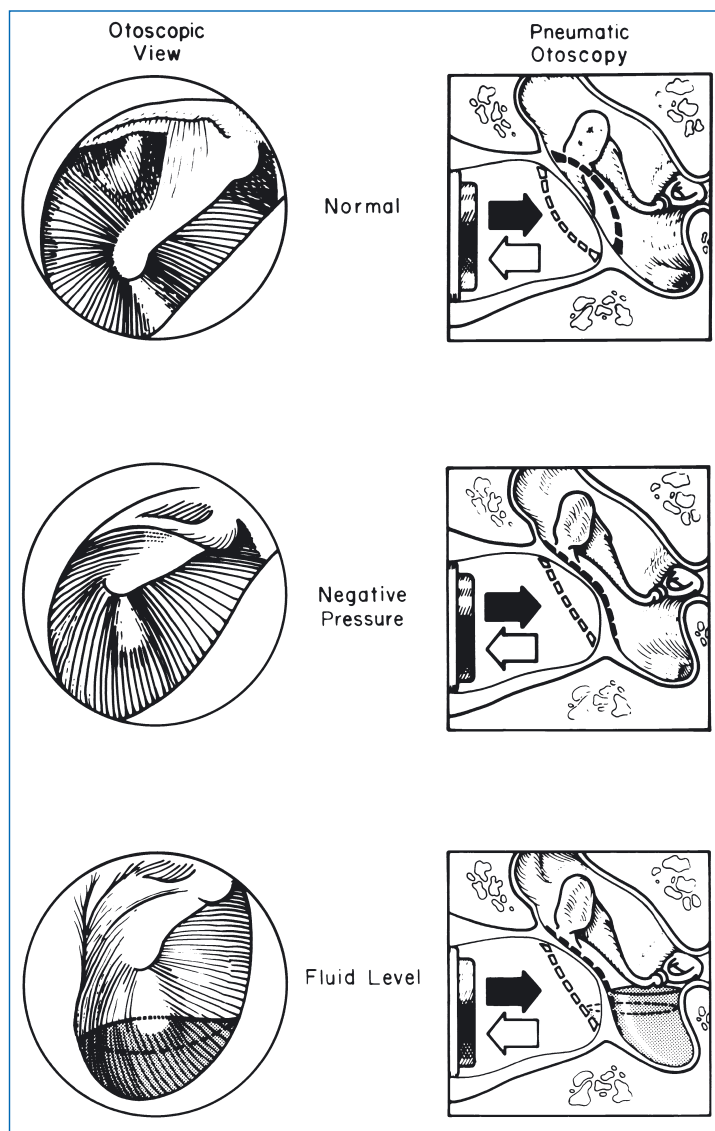


Fig. 16.2. Normal: The normal tympanic membrane has a neutral position and moves crisply with small amounts of applied or negative pressure. It is trans-lucent. Negative Pressure: The tympanic membrane is retracted toward the medial wall of the middle ear. It moves more on applied negative pressure than it does on positive pressure because it is already retracted. Fluid Level: The tympanic membrane is usually slightly retracted; fluid levels can be seen to move on application of positive or negative pressure. The color may vary with the thickness of the tympanic membrane and the color of the fluid

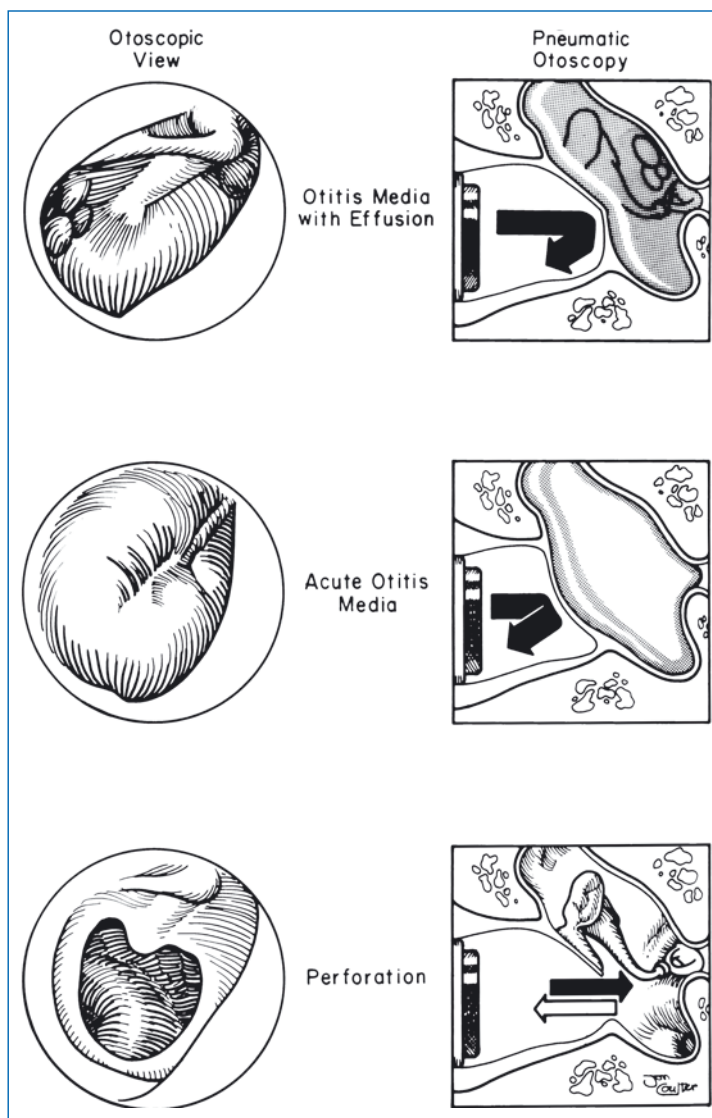
The operating microscope has been an important advance in visualization because it provides a powerful light source and enables the physician to clean the external canal and visualize the tympanic membrane. Recent optical advances, including the development of the rod and lens telescopes, have improved our ability to examine, photograph, and document the tympanic membrane structures.

Audiologic evaluation with pure-tone testing is usually used in children who can give voluntary responses and is important in serial evaluation of patients. Nonvoluntary responses obtained with auditory brain-stem testing have enabled us to demonstrate that young infants with cleft palates have a conductive hearing loss. Fria et al. [4] tested 23 infants and

demonstrated that 18 had a mild or moderate hearing loss in both ears and 4 had similar loss in one ear. Some of these were tested immediately postoperatively, and most of them had improved. This study confirms that even in the very young a hearing loss can be demonstrated.

Tympanometry or impedance testing is of great value in the cleft palate population, because it is very reliable in the detection of middle-ear effusion. In addition, this technique may be used to evaluate eustachian tube patency in the child who has an opening in the tympanic membrane, and there are some techniques that may be used on the intact tympanic membrane to show eustachian tube function.

Fig. 16.2. (continued) Otitis Media with Effusion: This is a frequent finding in the child with the cleft palate. The tympanic membrane is slightly retracted and there is very little movement on applied positive or negative pressure. The color may vary from amber to blue, reflecting the contents of the middle ear. Acute Otitis Media: The tympanic membrane may bulge outward and is usually opaque. There is little mobility, and the color may be yellow or red. Perforation: There is no movement of the tympanic membrane because the opening prevents pressure from building up in the external auditory canal. There maybe a small or large perforation or a tube may be present in the tympanic membrane and give similar results



16.1.2 Eustachian Tube

Dysfunction of the auditory tube has been postulated as a major cause of middle-ear disease in children with cleft palate. The major functions of the tube are ventilation and pressure equalization, drainage, and protection (Fig. 16.3). Although cleft palate may affect all of these functions, ventilation is probably the most important. The middle ear must be ventilated if the air-cell system is to develop in the temporal bone. Therefore, if the ear is filled with fluid during infancy, pneumatization will be affected. We have demonstrated that this occurs in children with cleft palate: a radiographic study of the temporal bone has revealed significant decrease in pneumatization of the system [5].

The auditory tube is located in the skull base, which includes an osseous portion and a neuromuscular portion (Fig. 16.4). The latter has a cartilaginous support which is attached to the tube musculature. The tensor veli palatini's primary function is to open the tube when it contracts. The lumen of the tube is lined with a mucosal epithelium. There has been much speculation regarding the relationship of the cranial base and the eustachian tube. It is thought that, when the cranial base is relatively flat and the tube is horizontal, the prevalence of otitis media increases in the cleft palate child and also in the normal population. Various alterations in the morphology of the cranial base have been described in the cleft palate patients, and it is obvious that these

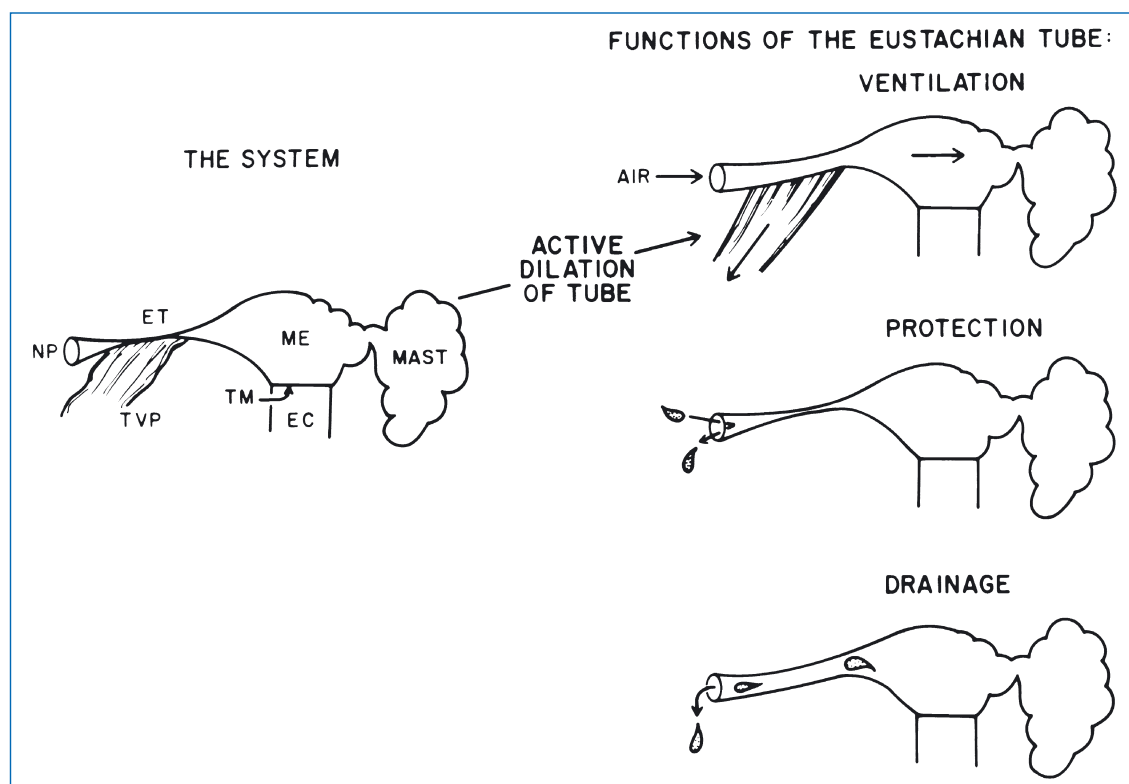


Fig. 16.3. The functions of the eustachian tube include ventilation, which provides pressure regulation, protection from secretions and sound, and drainage of secretions which may develop in the middle ear

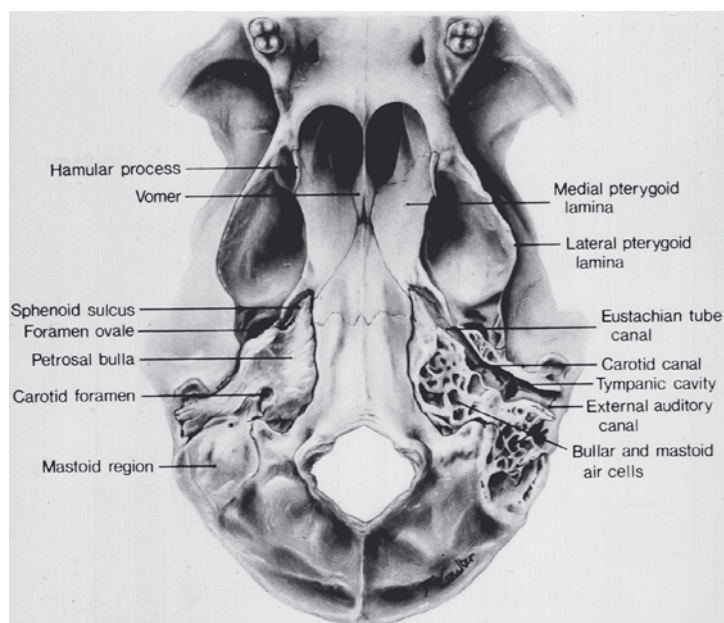


Fig. 16.4. The skull base showing its normal relationship to the bony eustachian tube. Alterations in the structure may result in a change of vectors of the musculature and malfunction

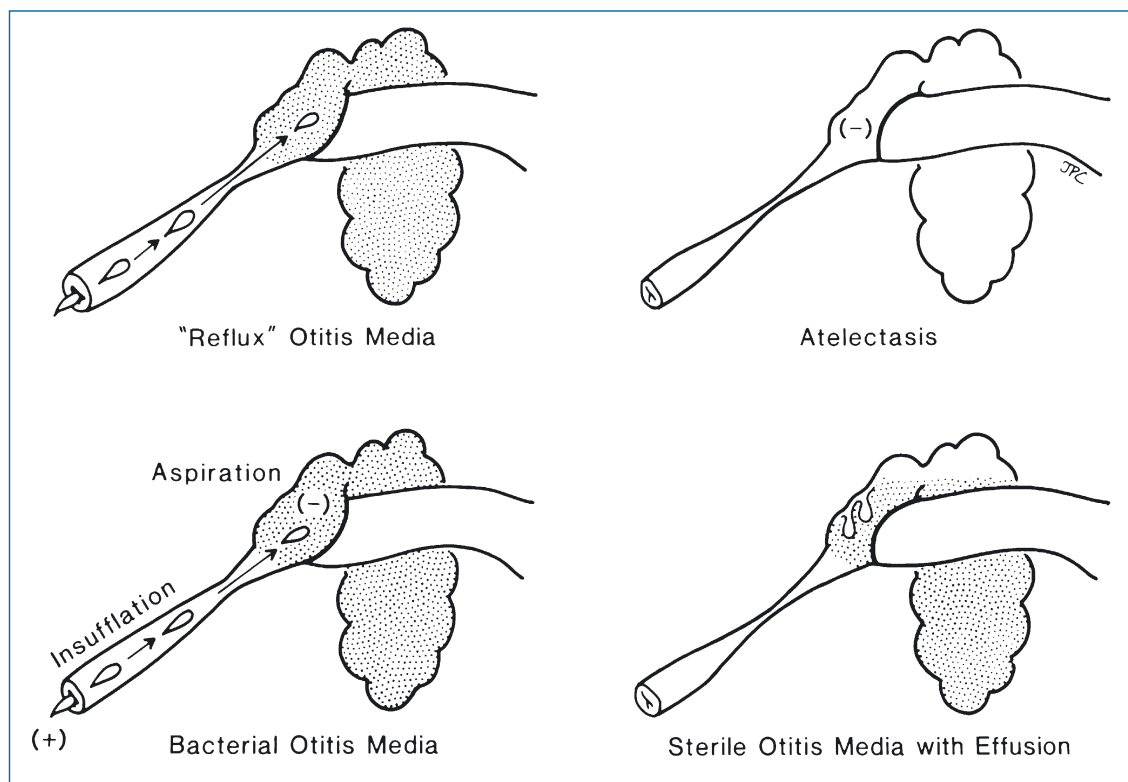


Fig. 16.5. The concepts of tubal malfunction that may result in otitis media – pus or bacteria in children with cleft palates may reflux into the tube and into the middle ear. The tube may be

“floppy,” and the middle ear can be insufflated. When the tube is obstructed, atelectasis may develop and a sterile effusion may occur, which subsequently may become infected

also may contribute to abnormal eustachian tube function [6].

Several mechanisms may result in middle-ear effusion due to tubal dysfunction. The tube may be obstructed or abnormally patent. In the event of tubal obstruction, the middle-ear space becomes atelectatic and can subsequently fill with sterile fluid (Fig. 16.5). This fluid may become infected if bacteria are insufflated into the middle ear. The tube also may be semipatulous or floppy, a condition thought to be prevalent in children, which makes the tube likely to dysfunction. In some instances, the tube is patent and secretions may have refluxed from the nasopharynx into the middle ear. We have seen children with cleft palate who have a perforation of the tympanic membrane, and have observed milk passing from the nasopharynx to the middle ear and out through the perforation to the external canal. Although these instances are rare, it does demonstrate the possibility of this anatomical connection and dysfunction.

16.1.3 Eustachian Tube Function Test

Because the eustachian tube is believed responsible for most of the middle-ear effusions in cleft palate patients, evaluation of its function is an important component of contemporary etiologic care. Researchers have expended considerable effort on developing methods of tubal evaluation. In general, the methods of evaluation fall into two categories: tests performed on the intact tympanic membrane and tests performed on the perforated tympanic membrane.

Tests on the intact tympanic membrane show changes in tympanometric measurements or demonstrate opening of the tube when sound or light passes through it. Sonometry utilizes sound, and some recent developments in Japan in which light-carrying bundles inserted up the tube can be detected in the external auditory canal have been reported [7].

Tests on the perforated tympanic membrane are most frequently used in clinical practice. Positive pressure is applied to the middle ear, and the pressure which causes the tube to open is ascertained; then positive pressure is applied below this threshold and

the patient is asked to swallow. This active swallowing should equalize the middle-ear pressure in about five swallows. Failure to do so indicates tubal malfunction.

The forced-response test is performed by opening the tube with a constant airflow through it and having the subjects swallow. In the abnormal subject, the pressure falls because the tube dilates; in the abnormal individual, there may be no active function or the tube may constrict, causing the pressure to elevate. These tests have been performed on a number of children with cleft palate. In infants with unrepaired clefts, there was a higher forced opening pressure than in infants with repaired clefts, and poor active dilating pressures [7]. The same group of children was tested following repair of the palate and was found to have improved in the passive functions, active function not being affected. However, a subgroup of children with very active ear disease demonstrated tubal constriction.

Experimental work on primates may be of value in understanding the relationship of the cleft to middle-ear effusion. Casselbrant [13], in studying the Rhesus monkey by producing a surgical cleft and then using the eustachian tube test previously discussed, showed that active tubal function was severely compromised by the cleft and that, with healing, the function was reversible.

A series of histopathological studies by Sando [8] on eight individuals with cleft palate produced several characteristic findings which suggest that the individual with cleft palate not only lacks anchorage of the tensor veli palatini muscle and the soft palate but also demonstrates abnormalities of the eustachian tube and its cartilage, as well as abnormal anatomical relationships of those structures to the tensor veli palatini.

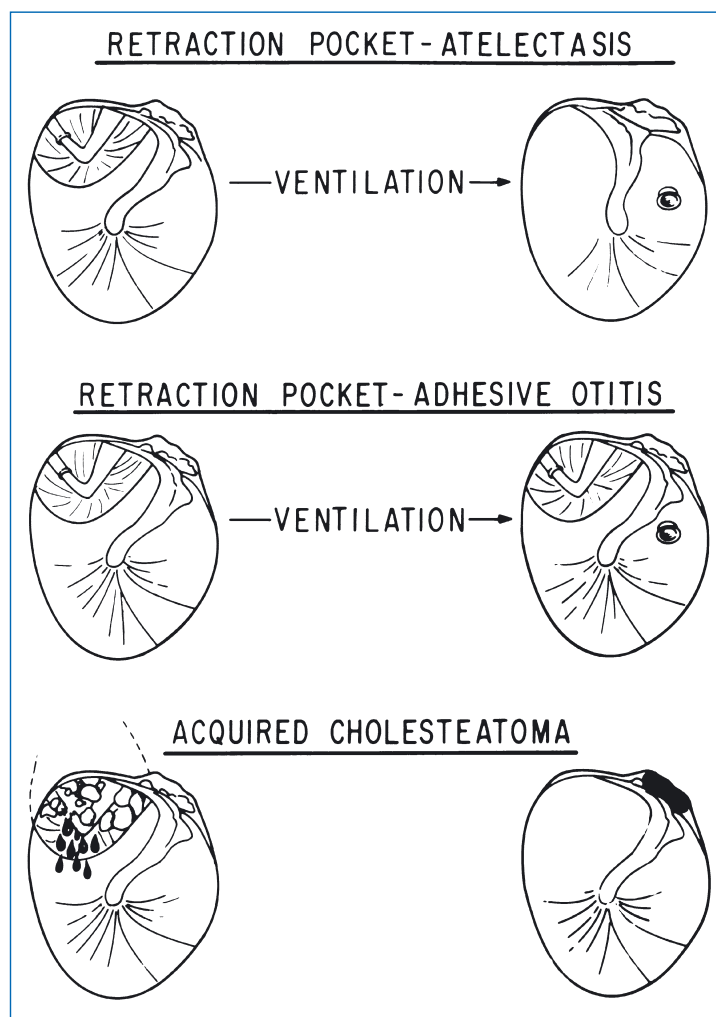


Fig. 16.6. The response of the tympanic membrane to ventilation of the middle ear. If there are no adhesions, an atelectatic tympanic membrane can recover. With adhesions there may be a permanent pocket. If it becomes filled with debris, a cholesteatoma can develop

16.2 The Sequelae of Middle-Ear Effusion in the Cleft Palate Child

Chronic otitis adversely affects any child's development. However, in the child who has a cleft palate, and therefore may have some difficulty with speech development, the long-term effect of middle-ear effusion or infections with suppuration obviously are more serious. Paradise et al. [9] addressed this question recently by comparing 24 closely matched pairs of children, one group with repaired palate clefts whose overall otologic treatment had been very aggressive and another group of children who had not had myringotomy with tube placement at an early age. The children were studied at ages 5–7 years, and both groups were similar in regard to physical and chronological findings. In both groups there was scarring of the tympanic membrane, and hearing was slightly affected. Although the articulation problems were essentially the same in the two groups, they were more substantial in the group that underwent later myringotomy. Verbal performance and IQ scores were essentially the same for both groups. From this study it was concluded that children with cleft palate do have some lasting adverse effects from otitis media during early life. However, it was not felt that this abnormality extended into cognitive, language, social, and emotional problems.

Anatomical alterations in the tympanic membrane and middle-ear structures of children with cleft palate remain a constant concern. Some children may develop tympanosclerosis, more frequently those who have grommets inserted, but have good hearing and basic structure [10]. Although there has been an increased interest in (in many instances aggressive) therapy utilizing myringotomy tube placement early in life, there still remains a group of children who develop unrelenting middle-ear disease and irreversible changes such as cholesteatoma. The response of the tympanic membrane to ventilation is shown in Fig. 16.6.

16.3 Craniofacial Anomalies and Communication

Communication involves primarily hearing and vision for sensory input and the vocal tract for output. In many instances, children with craniofacial anomalies have ocular defects that affect vision or such abnormal faces that it is difficult to establish eye contact; therefore visual communication may be poor. An example is Crouzon Syndrome, in which a shallow orbit results in a marked exophthalmos. Establishing eye contact may be difficult because the child's eyes may diverge and may have refractive errors.

The vocal tract may be abnormal with or without a cleft palate. In some disorders, such as severe Pierre Robin sequence, it may be necessary to perform a tracheotomy to provide an adequate airway, which may cause difficulty in communications. In children with microstomia or with a large tongue, oral communication will be altered. A small nasopharynx or nasal stenosis may result in hyponasality that interferes with communication.

Hearing loss that accompanies craniofacial anomalies may be congenital or acquired. The loss is congenital if it is present at birth and there is malformation of the external, middle, or inner ear. Hearing loss may be suspected if there is abnormal form or placement of the auricle. The presence of a microtia is obvious; however, abnormalities of the external auditory canal may be more cryptic. A small tortuous canal may become occluded with cerumen and a hearing loss may develop. All children with craniofacial anomalies should be suspected of having a hearing loss and should be followed carefully until their auditory acuity is established.

Abnormalities of the middle ear may involve the ossicular chain. This is especially prevalent in syndromes that affect the first and second branchial arches because these contribute to the development of the ossicles. An example is Treacher-Collins syndrome, a well-recognized entity in which ossicular chain abnormality has been described. The ossicles have a fetal configuration and do not conduct sound effectively.

Improved imaging methods have provided physicians with diagnostic methods of evaluating the ear; however, any child with a suspected abnormality of the external or middle ear should receive bone conduction amplification if there is a good possibility of conductive loss. In some children, reconstructive surgery is feasible but frequently must be delayed until later in childhood.

Acquired hearing loss is usually due to abnormal structure and functioning of the eustachian tube. This may be caused by abnormal cartilage and musculature of the tube or because the cranial base is abnormal and the rectors of the muscle are altered. Children with craniostenosis have a small nasopharynx, which also may result in tubal dysfunction.

The evaluation of the child with hearing loss has been described by Grundfast [11], and Sando [12] has described temporal bone findings in many of the syndromes. It is important to understand that diagnostic methods are available to evaluate almost all children regardless of age and that remediation of the hearing loss with amplification or surgery is usually possible. It is recognized that the earlier remediation is accomplished, the more satisfactory the outcome.

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17

Timing of Cleft Palate Closure Should Be Based on the Ratio of the Area of the Cleft to That of the Palatal Segments and Not on the Age Alone

SAMUEL BERKOWITZ

Diagnosis and surgical treatment planning in both medicine and dentistry are most frequently dependent on the patient's age, and nature and extent of the tissue's defect. In cleft lip and palate, the timing for surgically closing a cleft palate has been traditionally based solely on the age of the patient and the onset of speech (usually between 6 to 8 months) irrespective of the physical assets and defects of the affected tissue and not on the relative size of the palatal cleft defect to that of the surrounding palatal tissue.

The inability to develop quantitative diagnostic criteria to facilitate a differential diagnosis for proper treatment planning is partially due to cleft palate research programs having an insufficient number of investigative data obtained from serial maxillary and mandibular dental palatal casts of patients starting at birth and extending into adolescence as well as a proper instrument to measure the palatal cast's surface. The premise of this investigation is that data extrapolated from serial cast records will establish the relationship of the cleft defect to the palatal size and shape under the influence of corrective surgery.

Clinicians who do not have serial cast records have failed to appreciate the importance of cleft size/shape variations that exist within each cleft type at various ages, which may be crucial for making the proper decision as to when to surgically close the cleft space to avoid growth-inhibiting scarring.

With the advent of advanced technology to perform 3D spatial-temporal measurements and Cadcam computer software for in-depth analyses, extrapolated surface data from palatal casts can now be subjected to highly sophisticated quantitative analyses to perform differential diagnosis and treatment planning.

- **Hypothesis to be tested:** The hypothesis that a relationship exists between cleft size and palatal size to achieve good facial-palatal growth and speech will be tested, also, there is more than one physiological surgical procedure and the Hotz presurgical ortho-

pedic protocol does not increase palatal size in velocity.

17.1 Method and Material (Tables 17.1, 17.2)

Employing the palatal casts of 242 male and female individuals from eight institutions in the USA and western Europe, separate serial analyses were conducted of well-growing cases with excellent aesthetics, dental occlusion, and speech and a palatal control series of 17 nonpalatal cleft cases, to access the growth changes in size and velocity from birth through adolescence. These control cases consisted of various clefts of the lip and alveolus and/or soft palate but with no clefts in the hard palate. The various complete cleft lip and palate series were compared to this series. Malcom Johnson (personal communication) has confirmed that this control sample is an appropriate one for the palatal growth comparisons. In this group the mid-palatal suture, extending anteriorly to the incisal papilla at the anterior alveolar ridge, served as the medial border dividing the palate into right and left segments. With the exception of the excellent sample of cases from Goteborg, the vomer series had poor occlusion, facial aesthetics, and poor speech. This series was included in order to determine what timing and type of surgical procedure had produced favorable or unfavorable outcomes, and also whether the outcomes varied with the relationship of the size of the cleft defect to the size of the palatal segments medial to the alveolar ridges at time of surgery.

The participating institutions were selected on the basis of their excellent records and varied treatment protocols, and different racial, and mixed gender populations. The Miami sample of nonorthopedically treated cases was selected at random from a larger number of similarly treated cases.

Table 17.1. Number of Clinics and Patients Involved in Study

Clinics	Timing of surgery				Palatal closure procedure
	Lip Adhesion	Second Lip-Nose	2nd Alveolar Bone Graft	Hard and Soft Palate	
Miami	3 mos	6–8 mos	8–10 yrs	12–24 mos	Von Langenbeck + Vomer Flap
Illinois	2–4 mos	6–7 mos	Seldome	12–18 mos	Various procedure Wardil-Kilner, Von Langenbeck
Nijmegen	1–2 mos	udp 6 mos bdp 9 mos	10 yrs	SP 12–18 mos HP 6–9 yrs	Von Langenbeck
Goteborg	udp 1–2 mos	6–9 mos	9–11 yrs	SP 6–8 mos	Vomer Flap
Delayed	bdp 2–4 mos			HP 9–11 yrs	
Goteborg	2 mos	18–20 mos	average 14 yrs	2 mos	arterior vomer flap,
Vomer				9 mos	posterior push back
Northwestern	6–8 wks primary bone graft	6 mos		12 mos	intravelar – veloplasty

Twelve different groupings of cases were established depending on their institutional location and type of cleft (complete unilateral cleft lip and palate, CUCLP, and complete bilateral cleft lip and palate, CBCLP), with each group consisting of both males and females. The number and type of cases were as follows: University of Miami: 26 CUCLP, and 17 CBCLP; University of Illinois: 12 CUCLP; Northwestern University: 22 CUCLP; Nijmegen: 10 CBCLP; Goteborg-Delayed Closure: 24 CUCLP, 8 CBCLP; Goteborg-Vomer: 23 CUCLP, 10 CBCLP; Amsterdam: 26 CUCLP, 4 CBCLP; Rotterdam: 60 CUCLP. The statistical analyses of the CUCLP and CBCLP cases were carried out separately.

The Amsterdam and Rotterdam cases, which had been treated with the Hotz presurgical protocol (PP), were followed from birth for 48 months, permitting a determination of the PP on palatal growth effects differed due to presurgical orthopedics.

- *Analysis to be made:* Comparative effects of treatment of (1) the palates surface area's rate of change (velocity) and growth (size in 2 mm). (2) Size of the posterior cleft space and the velocity of its change. (3) Ratio of cleft size to the palate's size before and after surgery (total surface area). Each of the subsamples of cleft children will be compared to each other and to the age-appropriate control samples.
- *Data Extrapolation from the surface of palatal casts (Fig. 17.1):* A highly accurate, 3D, electromechanical palatal cast measuring instrument which gave a measurement error of less than 5% made possible a spatial-temporal (4D) analysis of palatal form and size changes, permitting an in-depth study of how the cleft space and palatal segments of each clinic's cases changed in relationship with each other over time. (Figs. 17.2a, 17.3)

- *Features to be measured and analyzed (Fig. 17.1–17.3):* The size of the palatal segments are measured serially starting at birth and divided into two treatment periods. The first period ends at surgery to close the palatal cleft. The second period is after palatal cleft closure: it includes adding the remaining cleft space with the changing size of the palatal segments (total surface area). In CUCLP: (Fig. 17.2a, b) The sizes of the palatal segments are limited laterally by the alveolar ridges, and anteriorly by a line connecting the most anterior point on the alveolar ridge (AC and AC1). Posteriorly a line is drawn from point gingival (P and P1) which are equivalent to the pterygomaxillary fissure seen on cephaloradiographs which marks the posterior limits of hard palate. When this transpalatal line makes contact with the cleft, points PC and PC1 are created. PC to PC1 measures posterior cleft space width. Medial border of the palatal segments (AC-PC, AC1-PC1) is limited by the cleft space. In CBCLP (Fig. 17.3) the premaxilla's surface area is limited anteriorly by its alveolar ridge (PML-PMR).

17.1.1 Method Used for Analyses

Cleft Subjects: The subjects had unequal numbers of observations and were observed at different ages. In order to compare the responses of the different groups, the data was recoded into new age intervals of 2, 8, 18, 36, 60, 84, 120, 168, and 192 months. The groups from Amsterdam-Rotterdam were observed only for 4 years and were coded as 2, 8, 18, and 48 months. Infrequently, especially in the early months, subjects would have two observations in a recoded age interval. When this happened, the mean values of the observations were used. The within-group variances

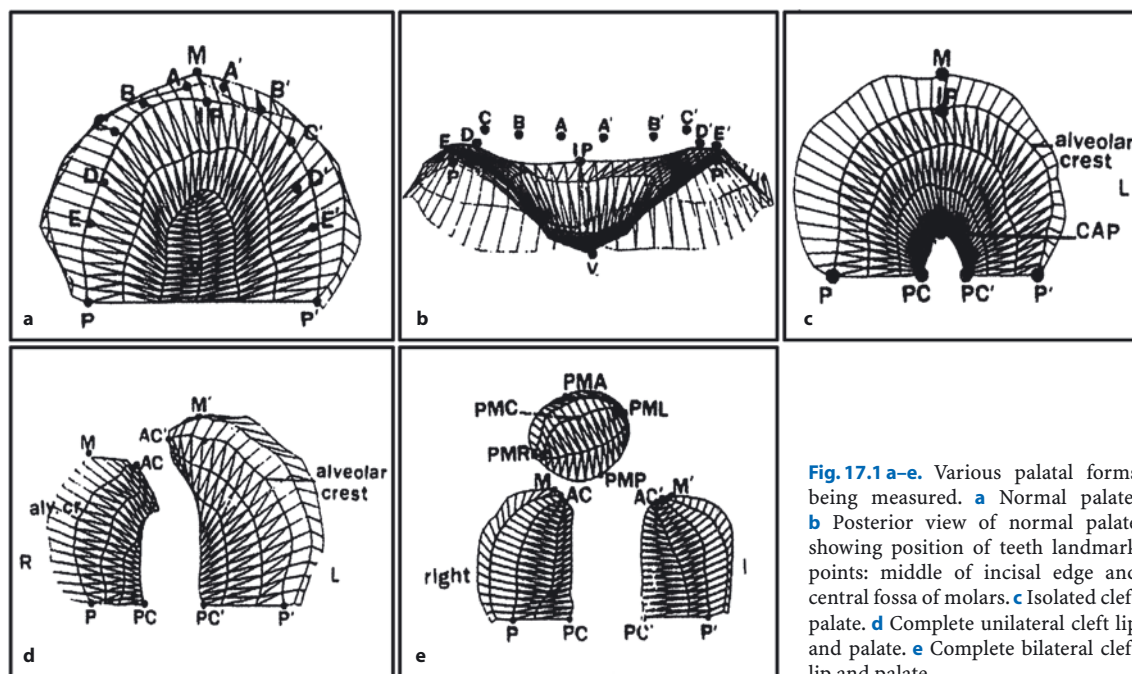


Fig. 17.1 a–e. Various palatal forms being measured. **a** Normal palate. **b** Posterior view of normal palate showing position of teeth landmark points: middle of incisal edge and central fossa of molars. **c** Isolated cleft palate. **d** Complete unilateral cleft lip and palate. **e** Complete bilateral cleft lip and palate

were compared during subsequent analyses to ensure that the equal-variance requirement of the Analysis of Variance was maintained and not biased by the use of mean values in some instances.

Controls: Serial casts from 17 patients who had only clefts of the lip and/or alveolus (CLA), with or without clefts of the soft palate (SP), or clefts of the lip and soft palate (CLA and SP) alone. All of these patients did not have a cleft in the hard palate. One case series had no cleft at all. The data from the control series was handled exactly as that from the cleft series.

Statistical Analysis: A preliminary analysis of palatal area was performed using a random effects model in the SAS Proc Mixed software. This preliminary analysis showed not only group differences, but, more importantly, heterogeneity in group responses over time (i.e., a significant Group x Time interaction). Because of the significant interaction, overall differences among groups or times are not directly interpretable. Consequently, the simple effects of differences between groups at each time point were investigated. The SAS Proc GLM software was used to test the fixed effects model among groups at each time, followed by a pair-wise comparison among mean values. At each time point it was sometimes possible to compare

among eight groups, which would lead to 28 comparisons (some time points had fewer groups because not all groups were observed at every time point). A strict control for multiple comparisons, say by the Bonferroni procedure, would severely decrease the power to detect differences. Therefore, the alpha level for each pair-wise test was set at 0.01. No statistical comparisons were made across time points but the consistency of the significant differences found in the pair-wise tests was noted. In this way, biologically consistent differences could be elucidated with sufficient power.

17.2 Treatment Protocols at Each of the Centers (Table 17.2)

17.2.1 Miami Craniofacial Anomalies Foundation, South Florida Cleft Palate Clinic (Figs. 17.1–17.3)

All of the lip and palate surgery was performed by D. Ralph Millard Jr., M.D., secondary alveolar bone graft by S.A. Wolfe, M.D., and all staged orthodontics procedures were performed by Samuel Berkowitz, D.D.S.

All cases studied from the participating institutions had similar graphs created.

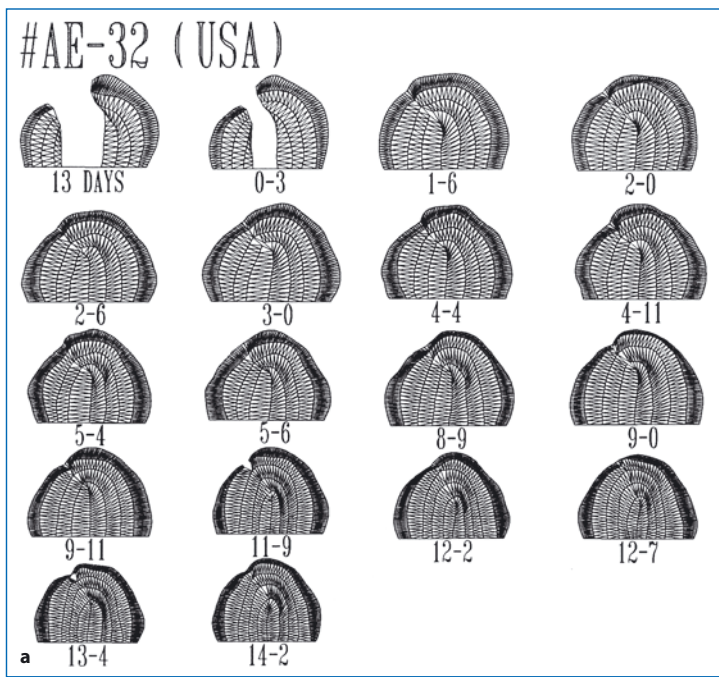


Fig. 17.2. a Twelve serial digital images of a complete unilateral cleft lip and palate from 13 days of age to 14 years and 2 months of age. The 2D images generated from 3D casts, are accurate in form but are not related in size to each other. The image at birth (13 days) is followed by (0.3 and 1.6) that had undergone molding as a result of muscular contractive forces created by lip adhesion at 14 months of age. Palatal cleft closure using a von Langenbeck with modified vomer flap was performed at 14 months of age. A secondary alveolar bone graft was placed at 8 years and 3 months of age after some minor orthodontics to align the anterior teeth. No presurgical orthopedics was performed

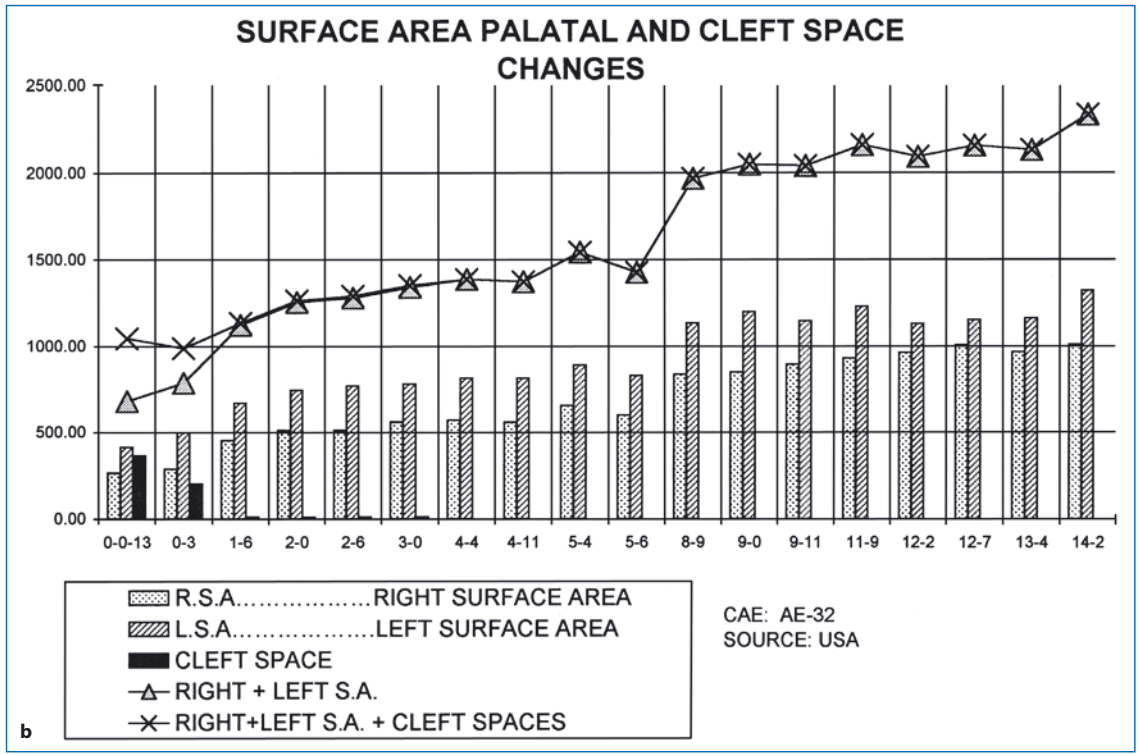


Fig. 17.2. b Graph depicting serial palatal 3D bony growth changes and the size of the cleft space of case 2A. The combined surface areas of the large and small palatal segments changed from 679 mm² at 13 days to 1121 mm² at 1 year 6 months (an increase of 39.5%) at the time of palatal surgery. The cleft space reduced from 365 mm² to 202 mm² at 3 months due to molding. Additional molding and palatal growth further reduced the

cleft space prior to palatal surgical closure. Palatal growth occurred gradually up to 5 years and 6 months. The continued marked increase in palatal sizes up to 10 years, 7 months reflects posterior palatal growth to accommodate the developing molars. This posterior palatal area appears not to be affected by the palatal surgery

Table 17.2. Treatment Protocol and Sequence

Controls = (Miami)	Soft Palate + 5	Lip/Alveous + 5	Soft Palate Lip Alveous 7
Clinics	CBCLP	CUCLP	Presurgical Orthopedics
Miami	17	26	None
Illinois	0	12	None
Nijmegen	10	0	Hotz 3–4 wk for 2–3 yr
Goteborg Delayed	8	24	1 wk to 1–1.5 yr
Goteborg Vomer	10	23	1 wk
Northwestern	0	23	Posterior 1/2 palate

Fig. 17.3. Digitized serial 2D cast images of a one side incomplete and the opposite side complete bilateral cleft lip and palate. The serial form cast's serial changes are accurate but not so as it relates to their relative size. At 0-1 (1 month) palatal segments are laterally displaced. At 0-4 (4 months), this image shows medial movement (molding) of the palatal segments, as a result of lip adhesion. The premaxilla is positioned forward of the lateral palatal segments with marked closure of the cleft space. At 0-8 and 1-1, these images show further spontaneous closure of the palatal cleft space as a result of palatal growth. At 1-4 palatal closure was performed 1 month earlier using a von Langenbeck with a modified vomer flap. Note that the following cast images show the gradual incorporation of the premaxilla within the palatal arch with orthodontic treatment



17.3 Results – Comparison of Total Surface Area in Unilateral Cases

The mean values, standard errors, and sample sizes for the unilateral data are shown in Table 17.3 and Fig. 17.2a,b. From Table 17.3 it appears that all other groups are below the Controls and that the Goteborg-Vomer group and the Northwestern group behave somewhat differently from the rest. Both are lower. The results of the statistical comparisons are shown in various Tables which are set up as a matrix to be read as follows: In the cell defined by the intersection of a row for one group and the column for another group

the times where the two groups are significantly different are indicated. As an example, following the Miami row and the Goteborg-V column, we see that these two groups had significantly different total areas at 36, 60, and 84 months. That this is reasonable can be determined by inspecting Fig. 17.6, Unilateral Cleft Lip and Palate Data, or looking at the mean values in Table 17.4.

The most striking feature in Tables 17.3 and 17.4 is that all of the groups differ from Controls over time. (*see note) The differences decrease over time due to growth changes in the posterior molar area. This growth area is associated with the developing perma-

Table 17.3. Total Palate Surface Area (Mean $\pm$ Std. Error, Sample Size) of Unilateral Clefts by Data Source and Age in Months

Source	Months							
	2	8	18	36	60	84	120	192
Amsterdam	817 $\pm$ 19 (33)	1003 $\pm$ 25 (33)	1157 $\pm$ 22 (31)	1396 $\pm$ 29 ^a (33)				
Miami	811 $\pm$ 22 (28)	1001 $\pm$ 24 (22)	1142 $\pm$ 25 (24)	1237 $\pm$ 23 (30)	1402 $\pm$ 32 (30)	1616 $\pm$ 40 (28)	1806 $\pm$ 52 (28)	2144 $\pm$ 75 (18)
Controls	930 $\pm$ 34 (11)	1066 $\pm$ 34 (13)	1283 $\pm$ 40 (12)	1496 $\pm$ 36 (14)	1726 $\pm$ 57 (15)	1881 $\pm$ 65 (17)	2060 $\pm$ 78 (12)	2337 $\pm$ 123 (7)
Goteberg (D)	882 $\pm$ 27 (24)	986 $\pm$ 21 (22)	1126 $\pm$ 38 (18)	1220 $\pm$ 33 (21)	1384 $\pm$ 37 (20)	1599 $\pm$ 45 (20)	1830 $\pm$ 50 (24)	2248 $\pm$ 46 (23)
Goteberg (V)	842 $\pm$ 21 (23)	1013 $\pm$ 28 (22)	1052 $\pm$ 26 (19)	1109 $\pm$ 28 (20)	1189 $\pm$ 32 (20)	1406 $\pm$ 45 (22)	1717 $\pm$ 38 (23)	2119 $\pm$ 57 (21)
Illinois	892 $\pm$ 31 (11)	1012 $\pm$ 49 (11)	1108 $\pm$ 32 (19)	1242 $\pm$ 38 (17)	1309 $\pm$ 53 (11)	1681 $\pm$ 52 (9)	1889 $\pm$ 72 (10)	2293 $\pm$ 61 (9)
Northwestern	730 $\pm$ 21 (28)	999 $\pm$ 36 (12)	1084 $\pm$ 30 (11)	1126 $\pm$ 37 (9)	1316 $\pm$ 61 (7)	1480 $\pm$ 56 (6)	1653 $\pm$ 57 (21)	2265 $\pm$ 95 (6)
Rotterdam	779 $\pm$ 13 (60)	949 $\pm$ 18 (59)	1075 $\pm$ 23 (59)	1251 $\pm$ 25 ^a (59)				

^a Measurements made at 48 months.

Table 17.4. Statistically Significant Differences in Unilateral Total Surface Area ($p < 0.01$) at Specific Time Points by Sample Source

	Amsterdam	Miami	Controls	Goteberg D	Goteberg V	Illinois	Northwestern	Rotterdam
Amsterdam	–		2				2	
Miami		–	2, 18, 36, 60, 84, 120		36, 60, 84		2	
Controls	2	2, 18, 36, 60, 84, 120	–	18, 36, 60, 84	18, 36, 60, 84, 120	18, 36, 60	2, 36, 60, 84, 120	2, 8, 18, 36
Goteberg D			18, 36, 60, 84	–	36, 60	84	2	2
Goteberg V		36, 60, 84	18, 36, 60, 84, 120	36, 60	–	36, 84	2	
Illinois			18, 36, 60	84	36, 84	–	2	2
Northwestern	2	2	2, 36, 60, 84, 120	2	2	2	–	
Rotterdam			2, 8, 18, 36	2		2		–

nent molars. The other feature is that the Goteborg-Vomer group seems to be consistently low over the period of 36–84 months.

17.3.1 Growth Velocity in the Unilateral Cases (Fig. 17.5)

In order to compute the rate of growth (velocity) of Total Surface Area, the slope of the growth between time points was computed for each subject by dividing difference in area by the difference in time. The

slope was then assigned the midpoint of the two time observations for analysis and plotting.

The Northwestern and Illinois groups show the highest initial velocities, with Amsterdam, Miami, Goteborg-Vomer, and Rotterdam showing very similar intermediate velocities.

The Goteborg-Delayed group and the Controls show very similar but lower initial velocities. Interestingly, the Controls show a slight increase in velocity between week 8 and week 18. Between 18 and 60 months the Vomer group drops in velocity and stays constant. The other groups level off and stay

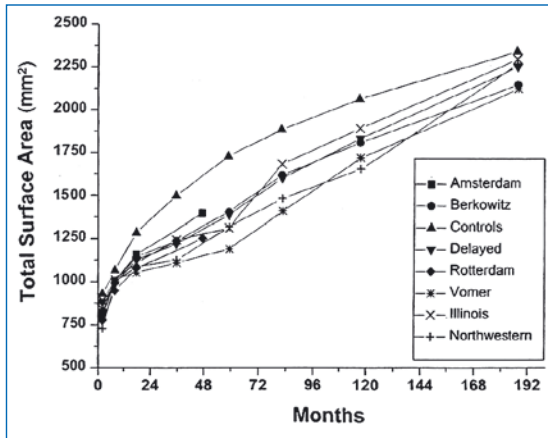


Fig. 17.4. Unilateral cleft palate data

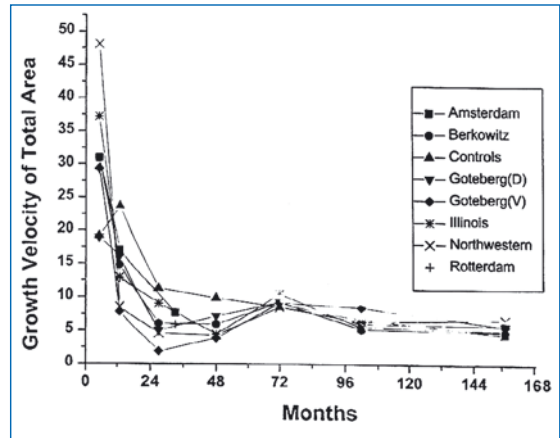


Fig. 17.5. Unilateral cleft palate data

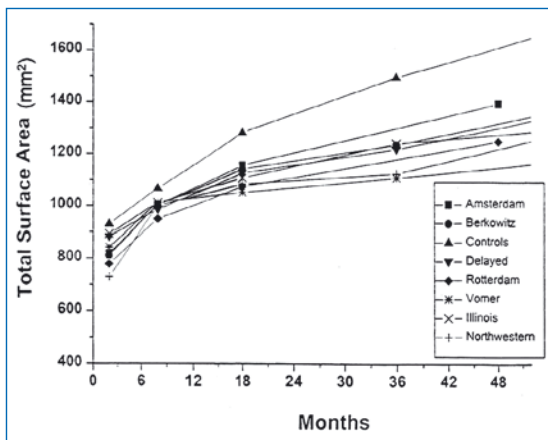


Fig. 17.6. Unilateral cleft palate data

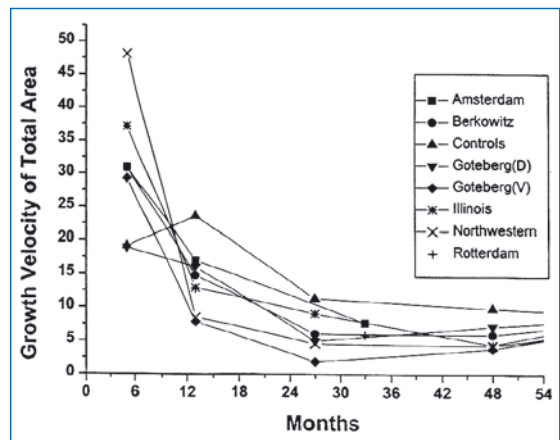


Fig. 17.7. Unilateral cleft palate data

approximately constant after 36 months. The plot of the mean velocities is shown in Fig. 17.4.

Some of the purposes of the Amsterdam and Rotterdam studies were to investigate growth velocity before surgery. The data on the velocity changes are plotted in an expanded graph in Figs. 17.5 and 17.7. This plot substantiates the observations made above.

These data show that the Controls have a substantially higher growth velocity than the other groups after the very early period. The Vomer group shows consistently low velocities. Interestingly, the Amsterdam and Rotterdam groups do not differ from Miami,

Goteborg-Delayed, Illinois, and Northwestern. The velocity data showed rapid growth early except for the Controls and the Goteborg-Delayed groups. During the period 8–18 months, the Amsterdam, Miami, Goteborg-Delayed, Illinois, and Rotterdam groups came together and showed constant growth thereafter. The Goteborg-Vomer group and the Northwestern group both showed a lower growth velocity than did the other groups. After the period of 8–18 months, the controls showed a consistently higher growth velocity.

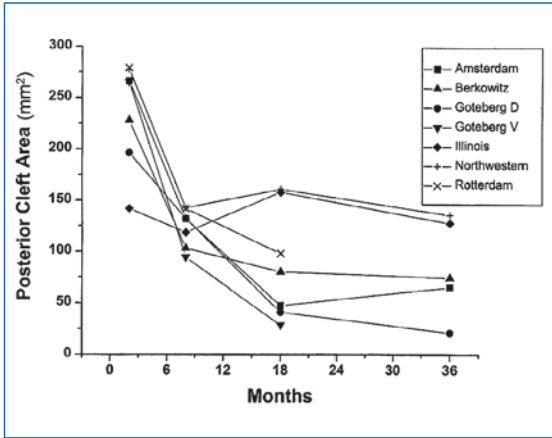


Fig. 17.8. Unilateral cleft palate data

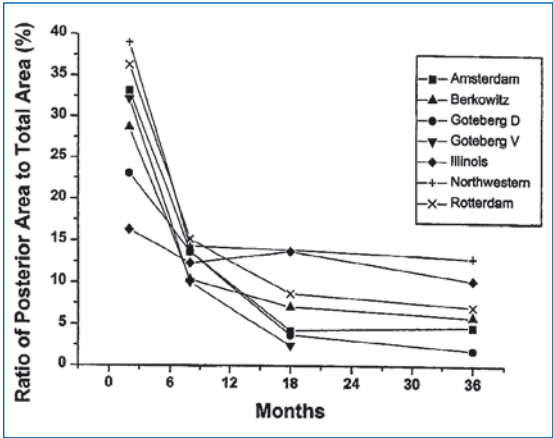


Fig. 17.9. Unilateral cleft palate data

17.3.2 Comparison of Unilateral Posterior Cleft Areas (Figs. 17.8, 17.9)

The outstanding feature of these data is that Illinois started out lower than the others and Northwestern started out higher than the others, but neither showed decreasing posterior cleft areas after 8 months. They actually increased in size to 18 months then slowly decreased. The other groups showed patterns similar to each other, becoming smaller in size at different rates and to different degrees.

17.3.3 Comparisons of the Ratio of Posterior Cleft Area to Total Surface Area in Unilateral Cases (Fig. 17.9)

At month 18 there were no significant differences between groups. These data have almost exactly the same pattern as the Posterior Cleft Space plots shown in Fig. 17.8.

17.3.3.1 Tracking of the Large and Small Segments in Unilateral Cases (Fig. 17.10 a–h)

The large and small segments tracked each other closely in all groups. Plots of the two segments for each unilateral group are found in the full-page plots. In early adulthood, although the data are sparse in this region, it appears that all four groups achieve similar total palate surface areas. The clinical decision to effect a relative early (18–24 months) closure of the unilateral cleft based on the Miami model appears to have had the same results as delaying the closure in the Göteborg-Delayed series until approximately 5–9 years of age. Both groups show a tendency for growth to start increasing at 5–6 years, coinciding with permanent molar development, and continue through adolescence and early adulthood.

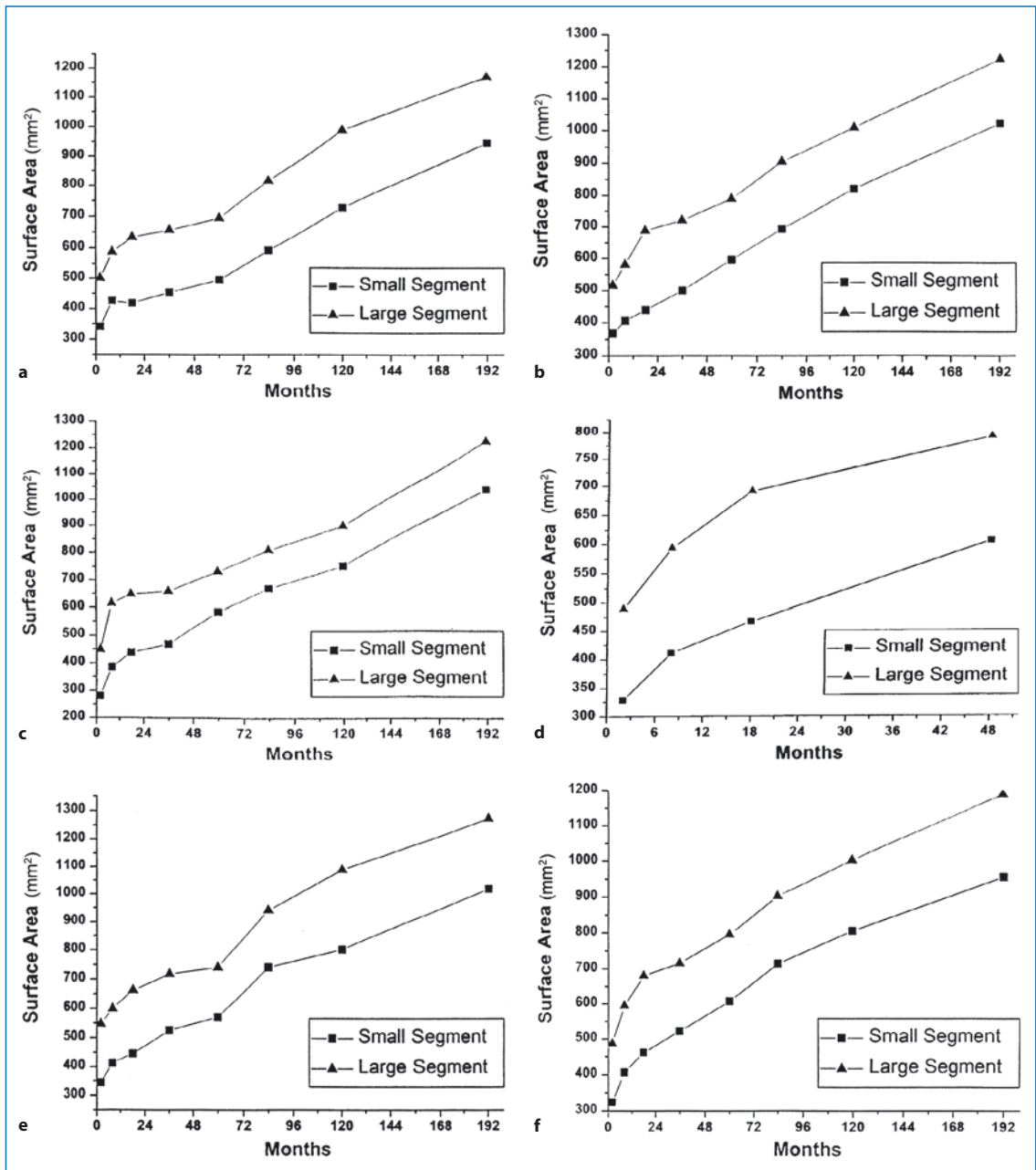


Fig. 17.10 a-h. Unilateral cleft palate data. **a** Goteborg Vomer Sample. **b** Goteborg Delayed Sample. **c** Northwestern Sample. **d** Amsterdam Sample. **e** Illinois Sample. **f** Miami Sample

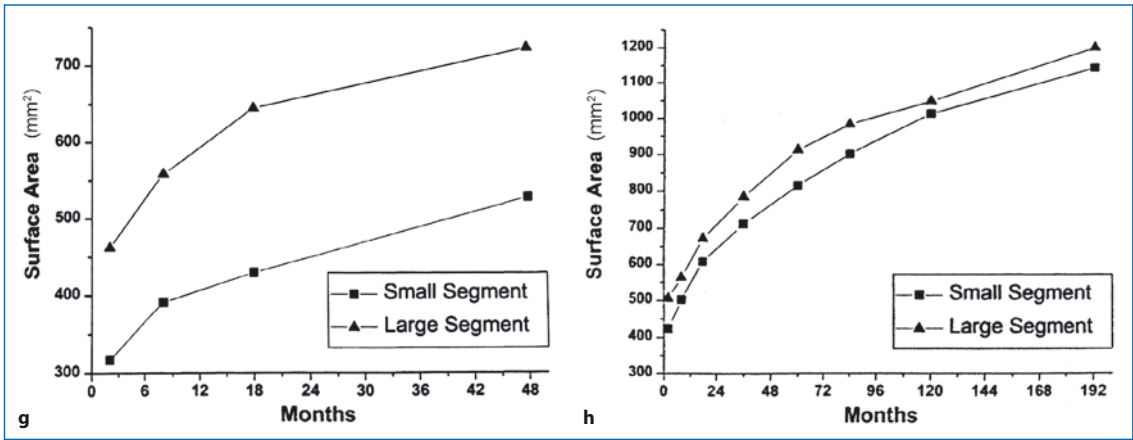


Fig. 17.10 a–h. (continued) g Rotterdam Sample. h Control Sample

Table 17.5. Surface Area Growth Velocity (Mean $\pm$ 1 Std. Error, Sample Size) of Unilateral Clefts by Data Source and Age in Months

Source	Months						
	5	13	27	48	72	102	156
Amsterdam	31.0 $\pm$ 2.9 (33)	17.0 $\pm$ 1.6 (34)	7.7 $\pm$ 0.7 ^a (31)				
Miami	30.9 $\pm$ 2.5 (22)	14.8 $\pm$ 1.9 (19)	6.1 $\pm$ 0.7 (23)	6.0 $\pm$ 0.6 (28)	9.3 $\pm$ 0.8 (26)	5.3 $\pm$ 0.7 (26)	4.9 $\pm$ 0.6 (16)
Controls	19.2 $\pm$ 5.7 (10)	23.7 $\pm$ 2.7 (12)	11.3 $\pm$ 1.4 (11)	9.9 $\pm$ 1.8 (13)	8.6 $\pm$ 1.3 (15)	6.8 $\pm$ 1.1 (12)	4.5 $\pm$ 0.5 (7)
Goteberg (D)	18.9 $\pm$ 3.2 (22)	16.1 $\pm$ 3.5 (17)	5.0 $\pm$ 1.4 (16)	7.2 $\pm$ 0.9 (17)	9.3 $\pm$ 1.0 (18)	6.1 $\pm$ 1.1 (20)	5.9 $\pm$ 0.6 (23)
Goteberg (V)	29.4 $\pm$ 3.1 (21)	7.8 $\pm$ 1.9 (18)	1.9 $\pm$ 0.8 (16)	3.9 $\pm$ 0.8 (17)	9.2 $\pm$ 1.2 (18)	8.5 $\pm$ 1.0 (22)	5.8 $\pm$ 0.6 (20)
Illinois	37.2 $\pm$ 8.6 (7)	13.0 $\pm$ 4.2 (10)	9.1 $\pm$ 2.6 (9)	4.5 $\pm$ 1.6 (6)	10.5 $\pm$ 5.3 (4)	5.6 $\pm$ 1.5 (5)	5.1 $\pm$ 1.5 (4)
Northwestern	48.1 $\pm$ 5.2 (7)	8.5 $\pm$ 2.7 (5)	4.6 $\pm$ 1.7 (4)	4.3 $\pm$ 0.7 (5)	8.6 $\pm$ 1.0 (3)	6.4 $\pm$ 0.7 (4)	6.8 $\pm$ 0.4 (4)
Rotterdam	29.2 $\pm$ 2.0 (50)	12.8 $\pm$ 1.3 (58)	5.9 $\pm$ 0.4 ^a (58)				

^a Estimate made at 33 month.

17.3.4 Comparisons of Surface Area in the Bilateral Cases (Table 17.5)

The statistical methodology for the bilateral comparisons was the same as that for the unilateral comparisons. The mean values for the bilateral cases are shown in Table 17.5. Clearly, the sample sizes are smaller than in the unilateral cases, but the standard errors reflect statistically stable observations. A plot

of these values is shown in Fig. 17.11. The most striking feature of these data is that the Control group and the Miami group are behaving similarly with respect to the other groups, although the Miami group shows lower growth until approximately 84 months. To show early differences more clearly, an expanded plot is shown in Figs. 17.13 and 17.14. These plots show clearly that the two Goteborg groups start off slowly.

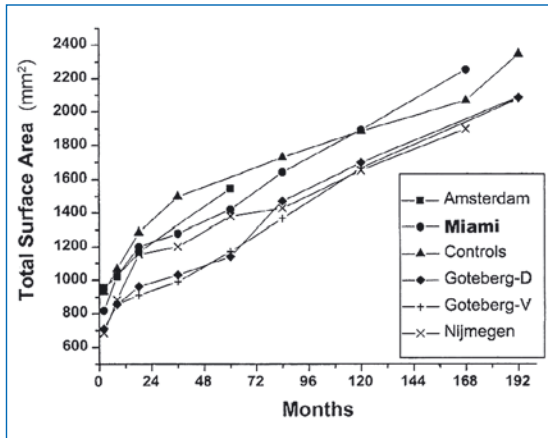


Fig. 17.11. Bilateral cleft palate data

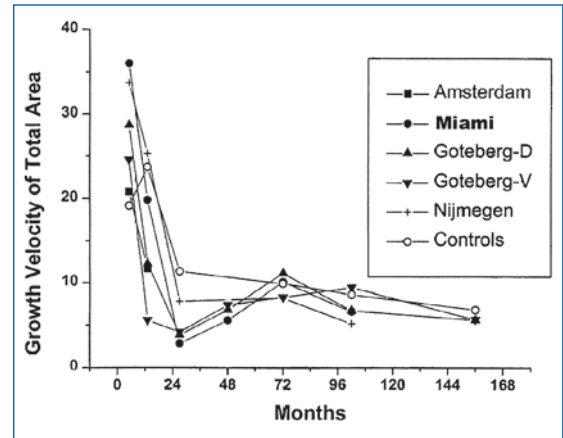


Fig. 17.12. Bilateral cleft palate data

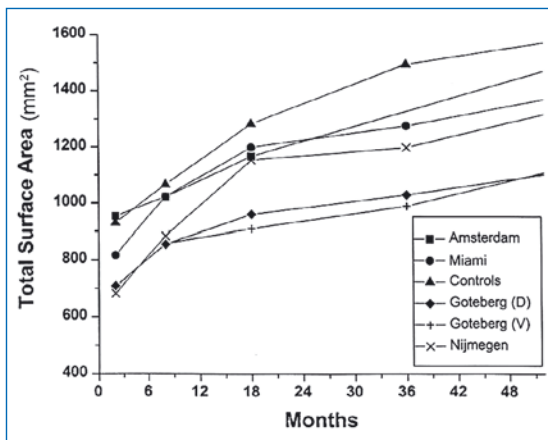


Fig. 17.13. Bilateral cleft palate data

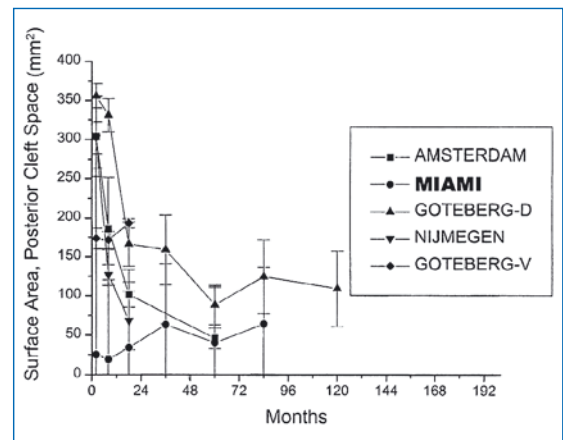


Fig. 17.14. Early growth velocity by group and time

17.3.4.1 Growth Velocity in the Bilateral Cases (Fig. 17.12)

The Miami and Nijmegen groups show the highest initial velocities, with Goteberg-Delayed and Goteberg-Vomer showing very similar intermediate velocities. The Amsterdam group and the Controls show very similar but lower initial velocities. Interesting, the Controls show a slight increase in velocity between week 8 and week 18.

Overall growth velocities are shown in Fig. 17.14. This plot substantiates the observations made above. Figure 17.12 shows that there is very little difference in growth velocities among the bilateral cases.

17.3.5 Comparison of Bilateral Posterior Cleft Areas (Fig. 17.15)

The posterior cleft areas up to 36 months are shown in Fig. 17.15. All the groups had similar patterns for the closure of the posterior cleft space.

17.3.6 Comparisons of the Ratio of Posterior to Total Surface Area in Bilateral Cases (Fig. 17.16)

At month 8 Nijmegen is significantly different from each of the other groups. There are no further statistically significant differences. These data have almost exactly the same pattern as the Posterior Cleft Space plots shown in Fig. 17.15.

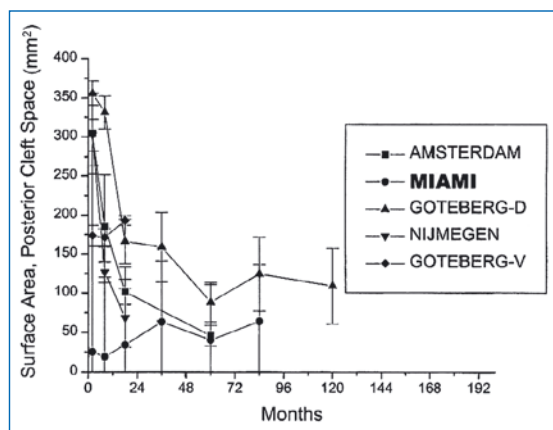


Fig. 17.15. Bilateral cleft palate data

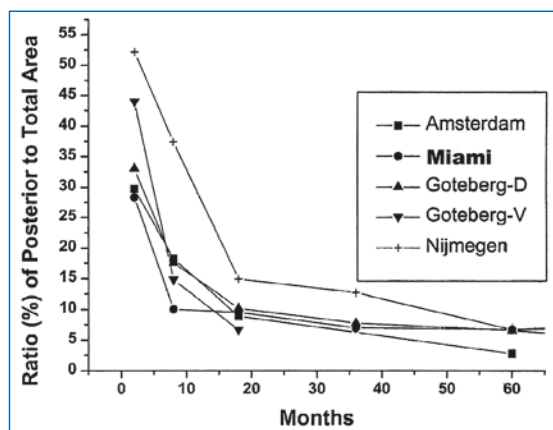


Fig. 17.16. Bilateral cleft palate data

Bilateral Cases: The Bilateral total surface growth curves revealed that both Goteborg groups grew slowly throughout the complete time period while the Nijmegen group showed slowed growth after 60 months. The Miami group showed a pattern similar to the Controls but with somewhat lower total growth between 18 and 84 months. The velocity profiles for the bilateral cases were very similar to those for the unilateral cases.

The closure of the posterior cleft spaces and the ratio of posterior cleft space to palatal surface area before surgery and to total surface area after surgery was also similar to that in the unilaterals. The only noteworthy feature was that the Nijmegen group had larger posterior cleft spaces and consequently higher ratios from month 3 through month 36. This may be due to the longer and more constant use of the presurgical orthopedic appliance which prevented the palatal segments from moving together (molding).

17.3.7 Conclusions for the Bilateral Series

The different practices showed similar results in palatal growth in that growth curves paralleled controls, but never entirely caught up. The only exception was that the Miami bilateral group did overtake the Controls. Early (18–24 months) or later closure had similar results.

17.3.8 Clinical Significance of the Results

- This study highlights that differences in palatal osteogenesis is reflected in differences in cleft space size at the same age at birth and for the next 12 months.
- The vomer series, where the palatal cleft was closed before 1 year followed by a velar pushback, showed that extensive pushback procedures interfered with palatal growth. This finding was reported by the clinic staff and was the reason for discontinuing this surgical protocol and changing to delayed closure without vomerine flaps and palatal pushbacks.
- The small modified vomer flap with a von Langenbeck procedure used in the Miami series did not cause palatal growth retardation or cause an excessive number of posterior crossbites.
- CBCLP cases appear to have more osteogenic deficiency than CUCLP cases at birth.
- All cleft palates are smaller than noncleft hard palates (controls).
- There is more than one physiological surgical procedure which can be used to close palatal clefts.
- In all cases the most common age for the smallest velocity of growth was between 18 to 24 months of age.
- The best time to close the palatal cleft is when palatal growth change has significantly slowed down so that cellular growth activity can proceed without interference.
- Physiological surgery (the procedure that interferes least with normal cellular activity) allows for catch-up palatal growth.
- It is now possible to mathematically determine the “best time” to close the palatal cleft, other than have it based solely on the age of the patient.
- It appears that the effect of cleft closure surgery involves only that part of the palate anterior to the first permanent molars. Subsequent palatal growth is necessary to accommodate the second and third molars and this area seems to grow independently of the effect from early palatal surgery.
- The lateral palatal segments in the CBCLP and CUCLP cases grow at a similar rate.

17.3.9 Discussion

Recent trends in the study of human biology generally and cleft palate research in particular exhibit an increasingly emphatic recognition of an important fact: that mass cross-sectional studies of large groups can be significantly less valuable than studies made of groups of single individuals over lengthy periods of time. Mass studies tend to smooth out significant individual differences and to obscure them; while consistent and prolonged serial studies focusing on individual areas tend to emphasize such variations by bringing them out in high relief. To date, investigating the growth of the palate has been studied only so far as it is displayed in the superficial soft structures overlying the bony palate.

All measurements in this paper, therefore, unless otherwise stated, refer to the soft structures of the palate and not to the skeletal palate. For purposes of convenience, whenever necessary, we shall hereafter in our series speak of the fleshy palate as distinguished from the bony palate.

The design of this study would have been better if it included test cases treated with the same palatal surgery but at 6–12 months as well as at 18–24 months of age. This comparative growth study would demonstrate the importance of using surgery based on the size of the cleft space relative to total palatal size and eliminate any resultant growth differences due to the type of surgery used. Unfortunately, we only had a small sample of these early closure cases. Since they developed excessive scarring with poor dental occlusion and midfacial growth, we did not accrue enough cases to perform valid statistical comparisons. Knowing that many organ systems in neonates grow very rapidly within the first 24 months, it was decided to use the same range of 12 to 24 months as the suspected ideal period for palatal cleft closure. A modified vomer flap was added to the von Langenbeck procedure to create a normal vault space. Since some cases had extremely large cleft palate spaces that reflected an excessively high degree of osteogenic deficiency, we focused on this factor rather than the age of the patient to avoid creating excessive scarring. The palatal growth velocity measurements showed that our reasoning as to when best to perform palatal surgery was supported by the subsequent analyses.

Surgical Goals: In 1938 Kilner listed the primary objectives of cleft palate treatment in order of importance to be speech, followed by chewing and aesthetics [1]. Unfortunately, this priority of goals still seems to be preferred by many plastic surgeons due to the influence of speech-language pathologists who fear the consequences of an open palate. Thus, they favor palatal cleft closure before 1 year of age. As a result, the

surgical history of cleft palate repair is replete with varied attempts to close the cleft space as if it was “a stagnant hole,” with minimal concern as to the surgical effects on palatal and facial growth [2].

Clinicians in all specialties have criticized the poor long-term aesthetic and dental occlusion results created by nonphysiological surgical procedures (as to type and timing) that did too much mucoperiosteal undermining too soon, creating excessive scarring.

Unfortunately, the speech argument for early palatal cleft closure before 12 months still prevails and with it the hunt for the “magical cut” that will answer all aesthetic problems at birth and in the future. Slaughter and Brodie [3], commenting on poor midfacial growth, stressed that reduction in blood supply and constriction by scars would jeopardize palatal growth, yet this message was disregarded. In addition, they stated that unwarranted trauma to hard and soft tissue, due to the fracturing of bone, and stripping of mucoperiosteum, would cause permanent damage to growth sites that were active until 5 years of age. There was no criticism of the timing of surgery, only the procedures being performed. Not having appropriate records, they could not relate the surgical outcome to the disproportionately small palatal segments to cleft size that existed before 1 year of age [4].

What surgical procedures to use and when to close the palatal cleft were questions that had no universally acceptable answers. While surgeons such as Veau [5] and Brophy [6] believed that early closure of the cleft palate improved speech development, there were many who took a contrary position. Koberg and Koblin [7] favored palatal cleft closure between 2 and 3 years of age to achieve both good midfacial growth and speech.

Delaying Palatal Closure: There were many European cleft palate clinics which emphasized in the 1960s to the present time the achievement of good midfacial, palatal development and speech. They recommended that cleft palate closure be postponed until the eruption of either the deciduous or as late as the permanent dentition [8–13].

Why Presurgical Orthopedics: The rationale of the four European centers in this study for using presurgical orthopedics within 2 weeks after birth is based on their speculation that this treatment may aid speech development and feeding. The Goteborg clinic uses the PSO appliance solely as an obturator to aid feeding. The Goteborg, Amsterdam, Rotterdam, and Nijmegen centers usually performed delayed, staged palatal closure between 5 and 9 years of age. Children’s Memorial Medical Center, Northwestern University Cleft Palate Institute, employs presurgical orthope-

dics with primary bone grafting while closing the palatal cleft at approximately 1 year of age. In some instances, PSO is also used to manipulate the lateral palatal segments to aid surgical closure of the lips and establish an alveolar butt relationship prior to primary bone grafting.

Advocates of delayed palatal closure (5+ years) wanted to avoid secondary malformations of the palate and severe deformities of the maxilla caused by scarring created by extensive mucoperiosteal undermining with transpositioning of the tissue. This occurred when surgery created exposed lateral areas of denuded bone due to stripping off of the overlying mucoperiosteum. Similar malformations were created in animals by Kremenak et al. [14].

To avoid the consequences of early surgery, Hotz [9] and Weil [12] advocated the use of an obturator in the interim until additional palatal growth occurred which reduced cleft space width. They believed that postponing palatal cleft closure would not jeopardize speech development. Bzoch [20] stated that early speech therapy, between 1 and 3 years of age, corrected early speech problems. Speech language-pathologists and surgeons have mistakenly disregarded the possibility that significant speech problems can be corrected with therapy.

Many benefits were claimed for the use of presurgical orthopedics, such as the stimulation of palatal growth, aiding speech development, and the reduction of middle ear disease. However, in a state-of-the-art report on oro-facial growth, no supporting literature was reported [15]. The same failure of having any supporting literature is in evidence even today.

The findings by the Amsterdam, Nijmegen, and Rotterdam prospective studies, which used presurgical orthopedics (PSOT) for 30-plus years, state that PSOT has a very limited effect on feeding, and have recently concluded that it has no lasting effect on palatal arch form. Therefore, the cost/benefit ratio may not warrant its further use [16]. They are beginning to question whether earlier palatal surgery may be warranted.

Berkowitz [17] believes the timing pendulums had swung too far to the opposite extremes: from early to very late closure, and is now swinging back again to early closure, between 6 to 12 months of age. Berkowitz [18] states that those who favor either extreme timing periods are still not focusing, as Berkowitz and Millard did, on the size of the cleft defect but continue to be fixated on the patient's age alone. Some surgeons have found a "middle of the road" treatment plan, around 12–18 months of age, but not on the size of the cleft space.

17.4 Good Speech Is Dependent on a Normal Palate

Sally Peterson-Falzone et al. [19] have written that malocclusion needs to be considered during the early speech learning years. She points out that the dental and orthodontic literature contains fairly consistent information regarding the effects of dental problems and malocclusions on speech.

A Need for Differential Diagnosis and Treatment Planning: A careful review of cleft palate surgical history makes it clear that a single mode of surgery based on age alone for all cases frequently results in severe palatal and midfacial deformities as well as poor speech development.

In general, this literature tells us that dental and occlusal problems are more likely to be causative factors in speech problems (1) when they occur in combination rather than singly, (2) when they are present during the speech-learning years as opposed to later years, and (3) when they influence the spatial relationship between the tip of the tongue and the incisors [18]. The literature also indicates that speech problems are fairly common when there is a restriction in the size of the palatal vault which is more apt to be found in Class III occlusions compared with Class II [18]. Children with clefts are obviously vulnerable to restriction in size of the palatal vault, and the possibility of Class III occlusions due to the presence of dental or occlusal problems (possibly several at one time) during the speech learning years. The question is: Will the speech problems diminish as the dentition or occlusion improves?

This statement convincingly acknowledges that good speech development is contingent on good tongue-teeth relationships within a normal vault space of proper volume.

This study has demonstrated that a scientific basis for selecting the best time to close the palatal cleft, in both CUCLP and CBCLP, is when the cleft space surface area is 10% or less of the surrounding palatal surface area bounded by the alveolar ridges.

Early palatal surgery (before 1 year of age in most instances) may not always jeopardize palatal and facial development provided conservative surgical methods are employed when the cleft space is sufficiently small.

The overarching thesis of this report favors consideration of the total emotional and physical health of the child with a cleft, based on the desired attainment of a cosmetically attractive face, and adequate dental function and respiration, as well as speech. Many sur-

gical, medical, and dental therapies may be necessary in the best-treated cases. As long as the surgeon individualizes the treatment plan, taking care to do no harm to growing structures, all goals are obtainable.

17.5 Conclusions

1. When the ratio of posterior cleft space to the total palatal surface area medial to the alveolar ridges is no more than 10%, it is the best time to surgically close the palatal cleft space. Therefore, one need not wait until 5–9 years of age to close the cleft space in order to maximize palatal growth.
2. Presurgical orthopedics does not stimulate palatal growth beyond its normal growth potential.
3. There is more than one physiological surgical procedure to achieve good palatal growth.
4. Extensive velar flaps with or without palatal push-back surgery is detrimental to palatal growth.

17.6 Participating Treatment Programs and Co-Investigators

Principal Investigator:

- Miami Craniofacial Anomalies Foundation, South Florida Cleft Palate Clinic, Samuel Berkowitz, D.D.S., M.S., F.I.C.D.

Co investigators:

- University of Miami, School of Medicine, Robert Duncan, MD.
- Center for Craniofacial Anomalies, University of Illinois College of Medicine, Carla Evans., D.D.S., D.M.Sc.
- Children's Memorial Medical Center, Northwestern University Cleft Palate Institute, Sheldon Rosenstein, D.D.S., M.S.D.
- Cleft Palate Center, Sahlgrenska University Hospital, Goteborg Sweden, Hans Friede, D.D.S., O.D.R.
- University Hospital of Nijmegen Cleft Palate Center, Anne Marie Kuijpers-Jagtman, D.D.S., PhD.
- Free University of Amsterdam Cleft Palate Center, Birte Prahl-Andersen, D.D.S., PhD.
- Academic Hospital (Dijkzigt/Sophia) Rotterdam Cleft Palate Center, M.L.M. Moberg, D.D.S.

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18 Neonatal Maxillary Orthopedics

SAMUEL BERKOWITZ

18.1 The Beginning

Looking back to the 1950s, it is hard to believe that in such a relatively recent period most cleft plate clinics in both Europe and the USA followed the lead of C. Kerr McNeil [1–3] a prosthodontist from Scotland who advocated presurgical neonatal maxillary orthopedics based on speculation rather than objective clinical data. Observing the dishd-in midfaces of many children with clefts, McNeil, not willing to accept that midfacial retrusion was due to retarded midfacial growth as a result of traumatic surgery, chose to adopt the thesis propounded by James Scott [4–6] an anatomist, who suggested that cleft palatal segments detached from the downward- and forward-growing nasal septum were deprived of their growth impetus, remaining small and deficient in mass and retruded within the face.

18.1.1 Stated Benefits of Presurgical Palatal Manipulation

McNeil [13] advocated that the maxillary segments distorted and displaced at birth be repositioned by a series of orthopedic appliances to produce a normal-appearing maxilla while reducing the cleft space in the alveolus and palate. This molding action, he theorized, would reduce nasal and lip distortion by bringing the palatal segments in closer relationships. McNeil further contended that the use of a plastic maxillary appliance, soon after birth, to align the palatal segments in an ideal relationship would correct the bony deficiency by stimulating palatal growth. McNeil misinterpreted the cause of the many retruded midfaces by seeing the condition as strictly a growth deficiency characteristic of children with cleft palate due to the palatal segments being detached from the vomer. He even went so far as to claim (without having examined serial casts to support his state-

ment) that the “stimulating growth” plate would eliminate almost all palatal clefts prior to palatal surgery. McNeil’s [1–3] plates were constructed from a series of modified plaster models in which the displacement of the palatal cleft segment was gradually reduced. Each successive plate when worn would gradually correct their position. These plates also carried “stimulator pads” that pressed gently on the palatal mucosa a short distance away from the margins of the palatal cleft. McNeil assumed that this pressure would stimulate growth of the underlying bone, thus reducing the width of the hard palate defect.

This activist philosophy of doing something at an early age found much favor with surgeons, because complete clefts were made to appear as incomplete clefts prior to lip surgery. Some advocates of this procedure have added unproven additional benefits, for example, improved speech and feeding and decreased middle ear disease.

Burston [7–9], an orthodontist from Liverpool, England became an early disciple of McNeil. Convinced that McNeil’s orthopedic concepts and methods were correct, Burston started a presurgical orthopedic ward at Children’s Hospital in Haswell, England. The protocol’s treatment principle was to establish early palatal arch symmetry in order to achieve normal dental function, palatal growth, normal respiration, chewing, and speech. Placing the palatal segments in an ideal relationship would stabilize the arch form and prevent growth retardation or collapse. It was anticipated that this procedure would permit the maxillary arch to develop normally and maintain its ideal relationship to the mandible. Sheldon Rosenstein [10–12], Nicholas Georgiade [13], T.D. Cronin [14, 15], Brauer et al. [16], Gruber [17], Manchester [18], Davies [19], Hotz [20–22], Nordin and Johnson [23], Monroe [24], Graf-Pinthus and Bettex [25], Graf and Bettex [26], Huddart [27–31], Robertson [32], Jolleys [33], McComb [34], Salyer [35], Huebener and Marsh [36], Stal et al. [37], and Nordhoff et al. [38] are

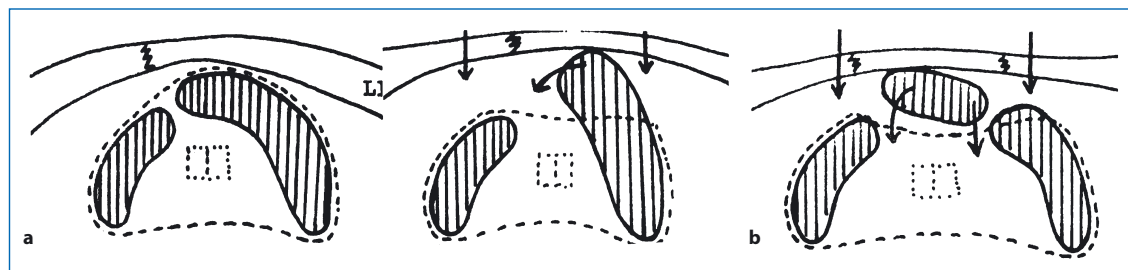


Fig. 18.1 a, b. Illustration of the Rosenstein appliance used in early maxillary orthopedics to bring the palatal segments into good approximation prior to performing a primary bone graft. It is inserted prior to lip surgery and is worn for 10–14 days after surgery. In most unilateral and bilateral clefts it is a pas-

sive appliance. In **a**, the unilateral cleft, the united lip molds the greater maxillary segment into a butt relationship with the lesser segment. In **b**, bilateral cleft, the appliance will hold the lateral alveolar segments apart until the premaxilla has been placed in close approximation. (For additional information, see [56])

ardent supporters of neonatal orthopedics in one form or another. Asher and Shaw [39] report that presurgical orthopedic treatment (PSOT) has been widespread and generally favored by British surgeons for more than 30 years and as of 1990 is still used in 39 of 45 centers surveyed, although its efficacy in terms of later outcome is widely doubted.

Many clinicians who followed McNeil's lead believed that every available means must be considered to prevent further development of maxillofacial deformities by constructing what were deemed better conditions for facial growth and occlusion and to maintain the corrected arch form. Norden et al. [40], Schmid et al. [41], Weil [42], Burston [7–9] Rosenstein [10–12] (Fig. 18.1), Rosenstein et al. [43], and Brauer et al. [16] in the 1950s and 1960s were the first to write glowing reports about such procedures. Georgiade [13, 44, 45], under the influence of Latham [46–49], advocated the use of pinned intraoral appliances. In the late 1970s, Millard, similarly motivated, followed Georgiade's lead. He also believed this orthopedic procedure would offer benefits not previously available, even though no reports on the success of this treatment had been published by the Georgiade camp. Millard started an orthopedic surgical treatment regimen for all types of lip/palate clefts which spanned the mid-1970s to the mid-1990s.

PSOT during the neonatal period has generated considerable and often heated debate about its utility and has divided many orthodontists and surgeons into different schools of thought. Some of the recommended treatments vary from passive holding appliances (see Fig. 18.1) [11] to the application of active orthopedic forces, such as Latham's pinned premaxillary retractive device.

Some proponents of early orthopedics selectively chose from a smorgasbord of unproven benefits. It was argued that such techniques would:

1. Establish normal tongue posture, aiding speech development
2. Aid in feeding
3. Help the surgeon in uniting the lip and closing the floor of the nose
4. Be a psychological help to the parents
5. Stimulate palatal bone growth
6. Reduce middle ear infections
7. Prevent the collapse of palatal segments, thereby reducing the need for future complex orthodontic treatment

18.2 Closing the Alveolar Cleft Space: Primary Bone Grafting

The obvious second step after aligning the palatal segments with presurgical orthopedics is to stabilize, if possible, the new geometric palatal relationships with bone. Those who advocated early palatal manipulation believed that the insertion of autogenous bone soon after birth and before the hard palate is closed was desirable.

The insertion of early bone grafts as a primary procedure in the treatment of cleft patients originated in Europe in the mid-1950s in conjunction with presurgical maxillary orthopedics. The graft, usually of autogenous rib or bone from the iliac crest, was inserted into the alveolar area via a buccal approach. The graft created a bony bridge into which, theoretically, new bone and teeth would migrate. By the 1970s many of those who initially advocated the procedure had abandoned it because their experience showed a negative effect on the growth of the maxilla and midface. From a study of a series of bone grafts, Jolleys and Robertson [50] concluded that the procedure resulted in no positive effects. On the contrary, they found reduced anteroposterior maxillary development with an

increased incidence of crossbites. Robinson and Wood [51], Friede and Johanson [52, 53], and Matthews et al. [54] also had strong doubts about the benefits of primary bone grafting. Peat [55] did not recommend bone grafting for stabilizing the maxillary palatal position after orthopedic treatment, but suggested that it might have a place as a secondary procedure after the age of 6 years.

18.2.1 The Kernahan-Rosenstein Procedure

Today's foremost proponents of neonatal maxillary orthopedic procedures with primary bone grafting are Kernahan and Rosenstein [56], who had been influenced by McNeil in 1954. In complete bilateral cleft lip and palate (CBCLP) children with a protruding premaxilla, Rosenstein uses a passive orthopedic appliance to cover the lateral palatal segments to permit the premaxilla to be molded back by an elastic extraoral facial strap (Fig. 18.1 a). In a complete unilateral cleft of the lip and palate (CUCLP), the acrylic appliance covers only the back two thirds of the larger palatal segment up to the alveolar ridge. The action of the surgically repaired lip will move this segment to an end-to-end relationship with the lesser segment. If the palatal segments overlap, Rosenstein uses a small jackscrew to create a gradual expansion of the contracted arch, prior to uniting the lip (Fig. 18.1). Bone grafting to the alveolus, using autogenous bone taken from the patient's sixth rib, is not done until the alveolar ridge is in an "optimum position" and then is performed to stabilize the segment. The appliance is worn for 6–8 weeks and may be used until just before closure of the cleft palate at approximately 18 months of age. They claim that the arches are stabilized within 4 to 5 months.

Although most cleft palate centers in the USA and Europe have abandoned primary bone grafting, Kernahan and Rosenstein still defend its use, claiming that the surgical procedure they use in placing the autogenous bone is significantly different from the procedures used by others and is the essential reason for their greater success-to-failure ratio.

Sadove (oral communication, 1990), attempting to duplicate the Kernahan-Rosenstein procedure, advises that, since none of the cases have reached maturity, it is still too early to file a final report.

18.2.2 Critical Review

No discussion of presurgical orthopedics and primary bone grafting would be complete without the critical review of the neonatal orthopedic procedures published by Samuel Pruzansky [57] in 1964. In his article,

the first paragraph sets the mood of Pruzansky's [57] dissent:

It is not simple or a lightly assumed task to write a brief challenging the rationale for presurgical orthopedics and bone grafting for infants with cleft lip and palate. The advocates are numerous and international ... Their battle cry is a cabalistic mumbo-jumbo invoking the mystic of embryology and growth and development. Their proposal to make things right and whole as early as possible seems sensible and has emotional appeal. Regrettably, and despite all this enthusiasm, what has been offered so far is a prolonged and costly manipulation and a surgery that is needless and sometimes barbaric.

Pruzansky strongly criticized the adoption of procedures whose results had not been adequately reviewed on the basis of objective diagnostic records of serial casts and cephaloradiographs to support their conclusions.

18.2.3 Long-term Results

Many of McNeil's original followers soon found the results of these procedures to be disappointing. Not only were additional orthodontics necessary, but it was suspected that presurgical orthopedics, as it was then practiced, might be causing more problems than had been anticipated, especially when the procedure incorporated primary bone grafting. Usually the autogenous bone graft is placed in the alveolar cleft space soon after the palatal segments are aligned at the time of closure of the cleft lip or shortly thereafter but before closure of the palate.

In 1978 Berkowitz [58] chaired a "State of the Art" report in cleft palate orofacial growth and dentistry. An international committee of 12 members reviewed the literature on the stated advantages of presurgical orthopedics. They found that most of the supposed benefits were not supported by the literature. As of this publication, the same conclusions hold because there is no new supporting literature.

A decade after Pruzansky's 1964 critique, many Europeans who had previously supported the procedure also changed their minds and condemned the results. Skoog [59] of Sweden, who had never favored the technique, wrote that presurgical neonatal orthopedics are both unnecessary and unhelpful, and suggested that well-performed lip repair would mold and reposition the maxillary segments into proper alignment.

Fara et al. [60], reporting on their long-term experience with neonatal maxillary orthopedics and primary bone grafting, concluded that it did not benefit

maxillofacial growth, and indeed had a detrimental effect.

Huddart [29, 30, 61], who knew McNeil personally, reports that McNeil put great emphasis on the use of stimulator plates as a means of reducing the hard palate defect, and stressed that they should be worn up to the time of palate repair at about 18 months of age. Huddart also observed that it was very difficult to get babies to wear these appliances consistently, especially beyond the age of 4–6 months.

Unfortunately, there are very few detailed reports of the results of presurgical treatment with active appliances that move palatal segments. In most cases, using passive appliances which control palatal molding action appears to be equally as ineffective on a long-term basis as employing active appliances which move palatal segments.

There are as many different types of passive plates as there are reasons for using them. Huddart's passive orthopedic appliance is a simple plastic plate that is inserted 24–48 h after birth. It obturates the cleft space but does not extend into the nasal cavity; it still, however, protects the underside of the nasal septum during feeding.

Huddart uses two adjustable wire wings that extend laterally from the corner of the mouth and lie on the cheeks to prevent the baby from swallowing the appliance. In CUCLP, the plate is utilized in conjunction with external elastic strappings to correct the anterior cleft defect by molding the premaxillary portion of the larger segment. Huddart reports a subsequent narrowing of the hard palatal cleft and believes that McNeil's type of treatment, which employs pressure pads, has an opposite effect, actually reducing the rate of tissue growth on the margins of the palatal cleft; however, this has not been proven. After years of using orthopedic appliances on cleft infants soon after birth, Huddart concluded that there appears to be no difference in occlusion between patients who receive PSOT and patients who do not.

Although many cleft palate treatment centers do not believe that orthopedic devices can stimulate palatal growth, they still advocate the use of neonatal maxillary orthopedics not only to aid in feeding but to bring the displaced palatal segments into proper position to facilitate the closing of the lip [62–68]. Some claim that the procedure is also psychologically beneficial to the parents at a critical time [69].

Friede [70] (Goteborg, Sweden), having gone through many changes in treatment philosophy, now argues that normalizing the arch form and postponing all palatal surgery to 5–6 years of age, when 80% of palatal growth is completed, can be highly beneficial. He suggests using external elastic forces with passive orthopedic appliances to retrude the premaxilla in CBCL/P and the premaxillary portion of the larger

segment in CUCL/P. The orthopedic appliance is utilized to maintain the arch form prior to the use of external elastic forces or lip repair. Although the Goteborg team claims excellent palate and facial development, superior to that achieved when the cleft space was closed at 1–2 years of age, they nevertheless are willing to accept the trade-off of increased management problems associated with a longer use of an obturator.

Even with all the ballyhooed but unsupported benefits, a small core of orthodontists still spoke out strongly against presurgical orthopedics (Pruzansky [57], Aduss [71], Subtelny [72], Berkowitz [58–73], Olin [74], Ross [75], Bishara [76], Mazaheri [77], Shaw [78], Bergland [79], Semb [80], and Friede [52, 53, 70]).

18.2.4 A Critique of Primary Bone Grafts

Cronin [71] was a strong supporter of early presurgical orthopedics and primary bone grafts in the 1960s. The many glowing reports by various European authors [77] were instrumental in encouraging him to use neonatal orthopedics with a primary bone graft to close the anterior palate followed by a vomer flap at 18 months and pushback repair of the hard and soft palates at 24 months of age. He also used secondary alveolar bone grafts in older patients, after the arch and occlusion had been corrected by orthodontics. At a second international symposium in 1969 held at the Cleft Palate Institute of the Northwestern University Dental School, Cronin [81] stated that the ideal of bridging the bony defect with a bone graft seemed logical. He listed the goals of the grafting procedure as: (1) to fix the alveolar segments together and equalize the growth; (2) to prevent collapse of the maxillary segments; (3) to stabilize the premaxilla in complete bilateral clefts; (4) to provide more bone for teeth adjacent to the cleft; and (5) to better support the ala and improve the contour of the cheek.

At this symposium, he reported that the bone grafting failed to prevent collapse but, to some degree, did stabilize the premaxilla in bilateral clefts. It also enabled teeth to erupt in the area of the lateral incisor and gave the ala support. At that time, he claimed that it was too early to judge the long-term effect on midfacial growth; however, he later reported that facial growth was retarded (personal communication, 1982). Most significantly Cronin [81], as cited by Cole [82], stated,

In looking over these cases through the years, it seems to me that the orthodontist has accomplished just as much at age 4 or 5 years as we have working with the infant, and that he has done it more easily and in a shorter period of time. My own view is that I would be

willing to wait until that period of time for this work to begin, after what I've seen.

A multicenter cephalometric study [73] of orthopedic versus nonorthopedic treatment in complete unilateral cleft lip and palate cases between 10 and 11 years of age involving 13 cleft palate centers showed no differences in the effect of presurgical orthopedics on the maxilla in either anteroposterior dimensions or growth patterns.

Berkowitz's [73] serial growth study of the use of the Latham-Millard pinned orthopedic appliance in complete unilateral cleft lip and palate case reported in 1995 at the American Cleft Palate-Craniofacial meeting in Tampa (Abstract 88) that, in the treated cases, a number of detrimental effects were related to use of anterior-directed forces on the cleft's lesser segment and the reactive posterior-medial-directed force against the premaxillary portion of the larger segment. The findings of this study showed that the cleft segment was usually displaced anteromedially, putting the buccal teeth in crossbite with the deciduous cuspid positioned in the lateral incisor space. The premaxillary portion of the larger palatal segment was displaced too far posteriorly and to the opposite side, causing the incisors to be positioned in an anterior crossbite relationship (Figs. 18.1–18.4). Berkowitz's observation of serial casts of neonatal maxillary orthopedic treated cases reported at this meeting that the position of the cleft's smaller segments within the maxillary arch was not stable, and it often moved medially into crossbite, even in the presence of a bone bridge across the cleft.

18.3 The Zurich Concept [84] (Fig. 18.2)

This approach is presented to demonstrate the treatment philosophy at a clinic where the main goal is to maximize palatal growth, and team members do not believe they are sacrificing speech development. They also believe that their PSOT has long-term utility.

Passive plates used in conjunction with delayed surgical procedures were introduced in Zurich, Switzerland in the 1960s after clinicians there had tried McNeil-type orthopedic procedures and found them wanting. The Zurich appliance is a combination of both soft and hard acrylic (Fig. 18.2). The plate penetrates into the nasal chamber to some extent, is worn continuously for about 16–18 months and is replaced every 6 months. Reduction of the gingival side of the plate every 3–8 weeks allows for palatal growth changes. A posterior extension to the top of the cleft uvula is a specific feature which Gnoinski [84] claims makes swallowing more normal. She stresses that the plate, while allowing for spontaneous development

without the tongue or other mechanical interference in the cleft, does not stimulate growth as claimed by McNeil [2–3], and Weil [42]. In connection with this sequence of treatment, the lip is united surgically at 6 months of age, the soft palate at 18 months, and the hard palate cleft is closed at between 4 and 5 years of age.

Gnoinski [84] lists the following benefits of the procedure: (1) it aids in feeding; (2) it maintains the width of the maxilla and lets the palate increase spontaneously in all three dimensions without impeding its growth potential; and (3) it permits the soft palate to grow to its maximum length prior to surgery.

18.4 The Netherlands Approach

PSOT in cleft lip and palate is provided in three centers in the Netherlands: Nijmegen, Amsterdam, and Rotterdam.

18.4.1 Unilateral Cleft Lip and Palate (UCLP) (Fig. 18.3)

A baby born with a cleft is referred to a Cleft Palate Center, preferably within 2 weeks after birth. The plastic surgeon and the orthodontist decide on the first treatment stages. In cases where the cleft is wide, PSOT usually is started immediately. The decision to start treatment is based purely on clinical experience. To fabricate the appliance, an impression of the upper jaw is made with an elastomeric material. A plate, consisting of compound of soft and hard acrylic, is carefully designed and fabricated to obturate the whole cleft laterally from right buccal vestibulum to the left (soft palate and alveolar ridge included) and placed in situ within a few days. The plate keeps the tongue out of the cleft. The support offered by the artificial alveolar ridge is thought to be important for speech development. The oral and nasal cavities are separated and nasal breathing is restored [75]. The plate is worn 24 h a day and held by suction and adhesion only.

Every 3 weeks the patient returns to the clinic to have the plate adjusted. Adjustments are made by grinding the plate to guide the segments during growth. The direction and total amount of growth are used to indicate when a new plate is necessary. A new plate is made the week before lip closure, which is done at approximately 5–6 months of age, to ensure that a well-fitting plate can be placed immediately after lip surgery. At this time, the alveolar cleft has narrowed considerably, which possibly facilitates lip closure [86]. In addition, because labial sounds appear in normal development at this age, a lip seal becomes essential.

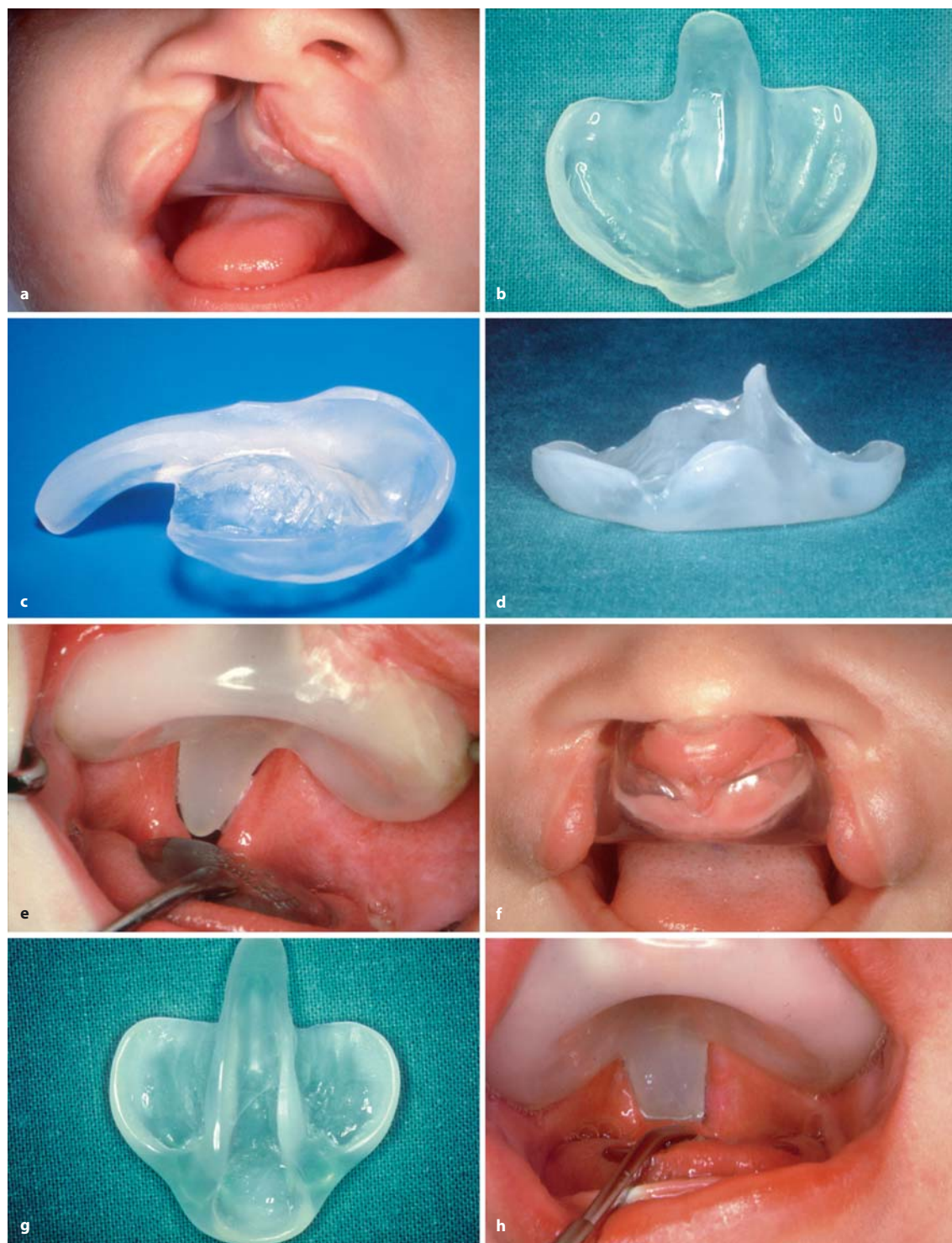


Fig. 18.2 a–h. PSOT appliances used in complete unilateral (a–e) and bilateral (f–h) cleft lip and palate. The palatal acrylic extends posteriorly to the uvulae. (Courtesy Wanda Gnoinski, Zurich University Dental Clinic, Cleft Palate Institute)

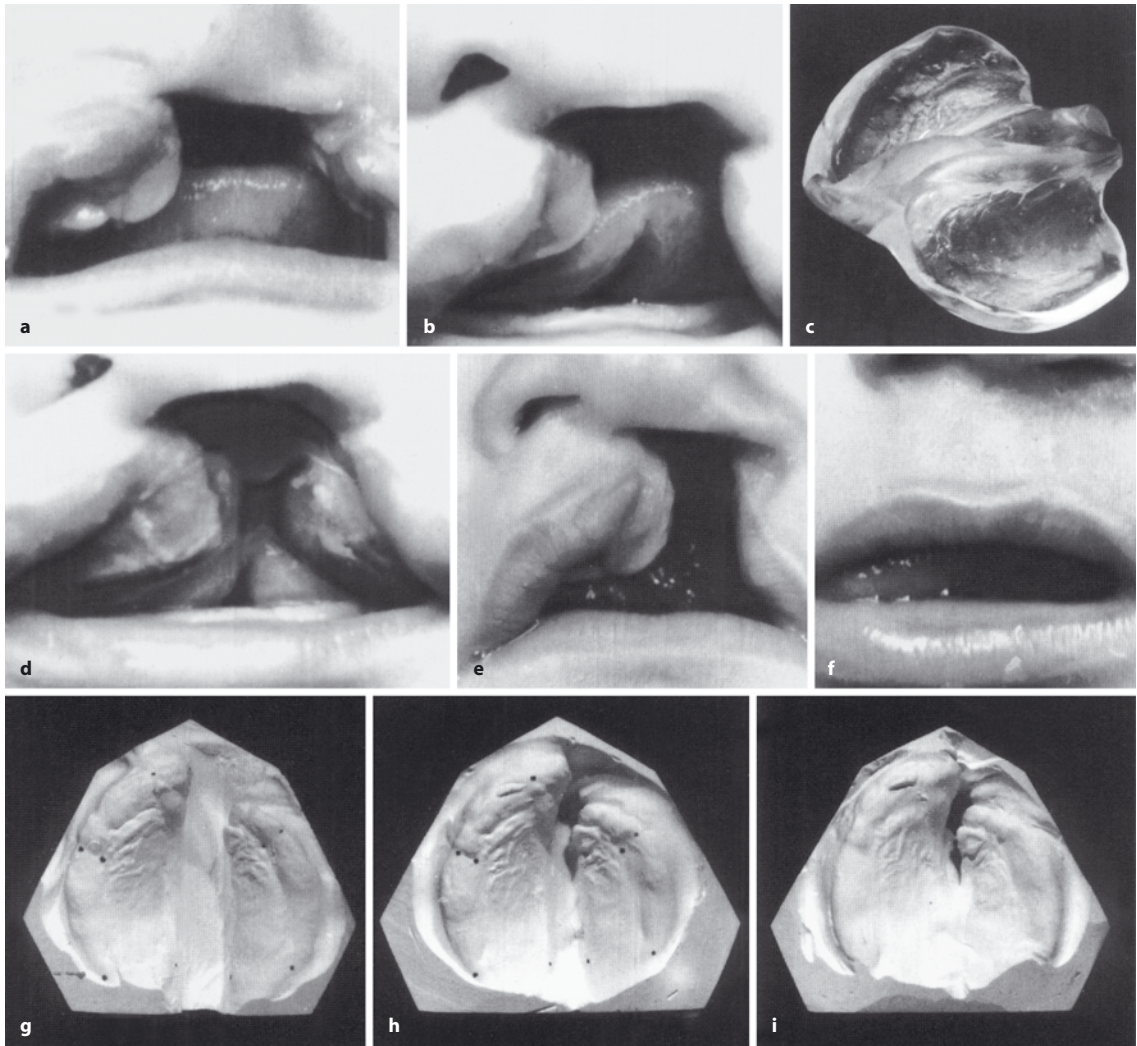


Fig. 18.3 a-i. PSOT appliance for a CUCLP utilized from birth to 11/1/42 at the University of Nijmegen (Courtesy of AM Kuijpers-Jagtman). **a** Lip and nose distortion at birth; **b** tongue posture within the cleft; **c** orthopedic appliance; **d** orthopedic

plate prevents the tongue from entering the cleft; **e** 15 weeks after PSOT and before lip closure; **f** 6 weeks after palate closure; **g** 17 months before soft palate closure; **h** at 14 months of age, before soft palate closure; **i** 8 weeks after lip closure

After lip closure (Millard rotation advancement), the new plate is inserted the same day to prevent positional changes of the maxillary segments, which are undesirable side effects of the operation. Then a two-stage palatal closure (modified von Langenbeck) is performed (i.e., the soft palate is closed at about 12–18 months and hard palate closure is done at 6–9 years of age) together with bone grafting of the alveolar cleft. The plate is worn until soft palate closure is performed (about 12–18 months of age). In the interim, the patient visits the clinic every 3 weeks. Later, when the growth velocity slows, visits are every 6 weeks. The plate is continually adjusted, as men-

tioned previously, taking into account the emerging deciduous teeth. Depending on maxillary growth, the plate is remade as necessary.

18.4.2 Bilateral Cleft Lip and Palate (BCLP) (Fig. 18.4)

The same procedures are performed, but extra-oral strapping is used. A one-stage lip closure (modified Manchester) is performed at about 6–9 months of age. Most surgeons believe, quite frankly, that one benefit of these procedures is to help fulfill the psychological

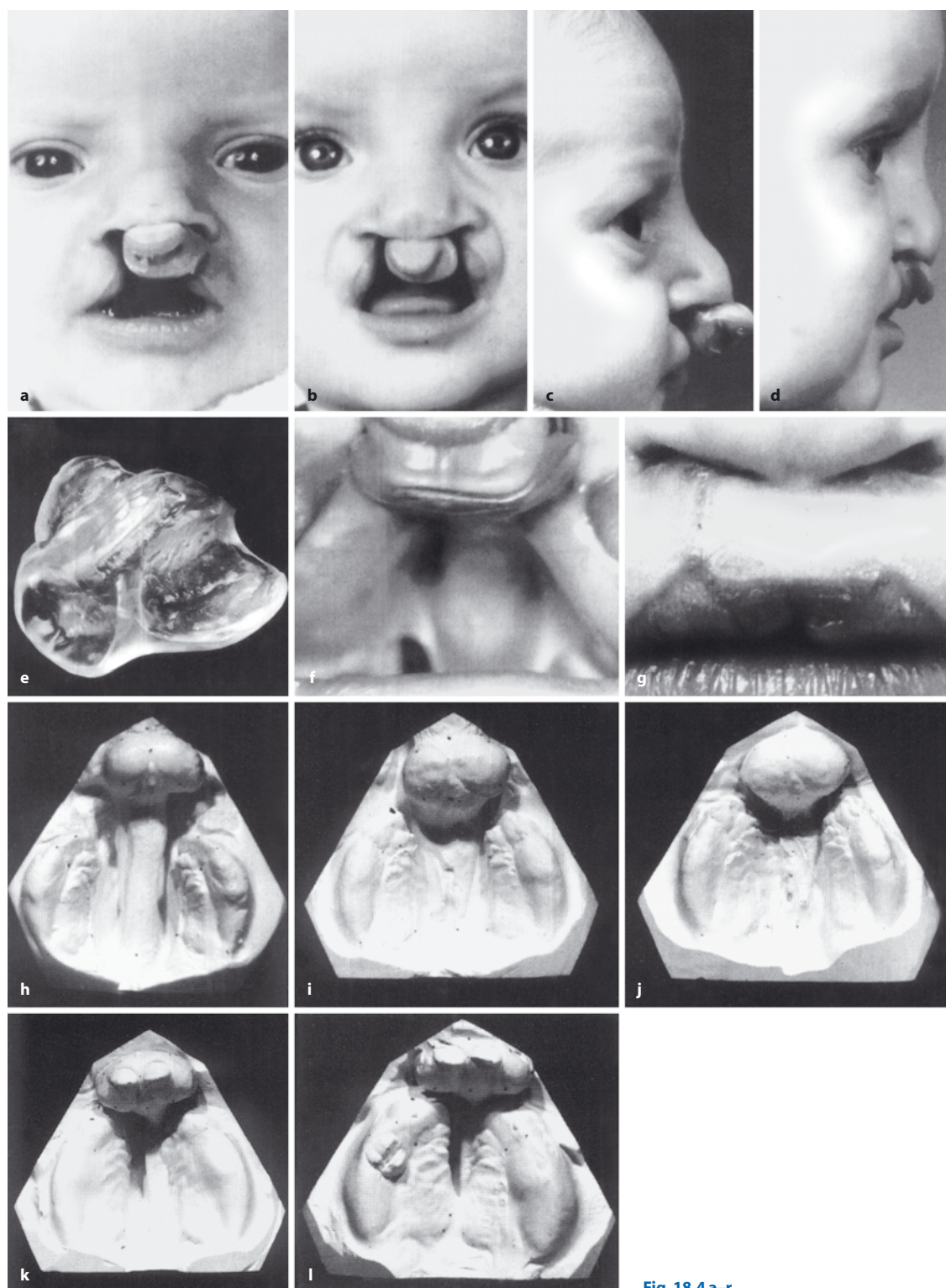
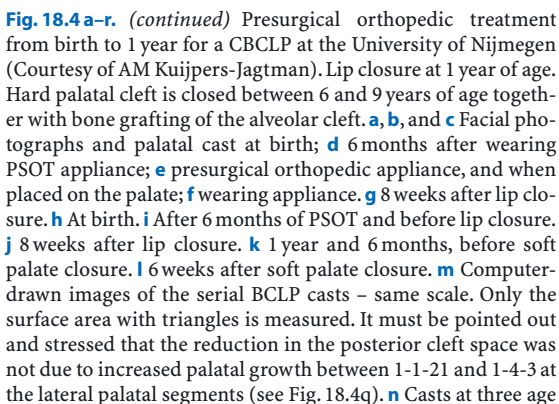


Fig. 18.4 a-r.



periods are superimposed on the baseline *P-P'* (the posterior limit of the hard palate drawn between postgingival points) and registered at *V* (vomer point on *P-P'* line). This shows the initial geometric changes in the alignment of the premaxilla to the vomer and the changes brought on by medial movement of the palatal segments, more on the right than the left side, and palatal growth increases with the reduction in the anterior and posterior cleft spaces. • Superimposing the casts from ages 3 months and 1-4-3 on rugae points shows the location of growth and molding changes of the palate. The premaxilla's protrusion was slightly reduced while the lateral palatal segments increased in size and moved medially (molding).

• Changes in the palatal slopes. *P-P'* line is the baseline for angular measurements. *AR*, alveolar ridge; *C*, medial limit of palatal segment in the cleft space. The angle between the two lines represents the slope

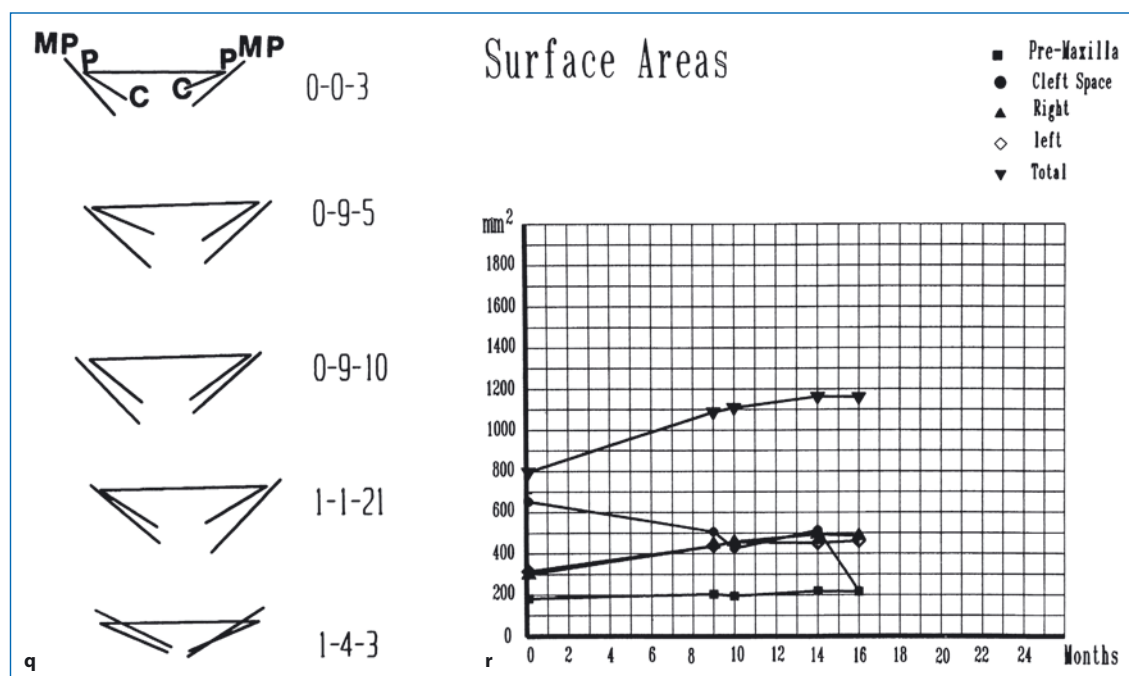


Fig. 18.4 a-r. (continued) q Serial changes of the palatal slopes and the cleft width between right and left segments. (MP, The slope at the middle of each palatal segment; P-P' line, The posterior limit of the hard palate; C, Medial limit of each palatal segment at the cleft) This series shows the gradual reduction in width of the cleft space. Between 1-1-21 and 1-4-3, the palatal slopes (MP) flatten, further reducing the cleft width. **r** Graph showing palatal surface area changes. The lines representing

the surface area of the right and left palatal segments, starting at 300 mm are superimposed and represent the growth curves of each palatal segment. The total surface area includes the pre-maxilla. The posterior cleft space at 14 months reduces from 650 to 500 mm. It then is markedly reduced to 200 mm at 16 months as a result of the flattening of the palatal vault. In 1 year, the palatal surface area shows an almost 50% increase, and the acceleration curve slows down

need of parents of children with clefts to feel that “something” is being done to help their child at birth, and feel that this is very important in maintaining harmonious family dynamics.

18.4.2.1 The Spread of PSOT Clinics

Rudolph and Margaret Hotz have attracted a following of eminent orthodontists who are more constrained in stating the benefits of their orthopedic procedures and have influenced the creation of PSOT clinics throughout Europe and the USA.

For an in-depth review of their treatment philosophy, the reader is referred to Hotz [20, 21], Hotz and Gnoinski [87], and Hotz et al. [88, 89].

Although clinics in Zurich and other European cities have accumulated an extensive number of lateral cephalometric films, photographs, and serial patient casts, they have not as yet proven that the Zurich-style orthopedic procedures have produced better long-term orofacial growth results than clinicians who have

not utilized neonatal maxillary orthopedics and have performed palatal closure between 1 and 3 years of age. Since this statement has been written in an earlier edition of *Cleft Lip and Palate*; the cleft palate clinics at Nijmegen, Amsterdam, and Rotterdam have reported on a prospective clinical trial testing the utility of PSOT using the Holtz treatment protocol.

18.4.2.2 Long-term Utility of PSOT

The hard fact is that, although many reasons have been offered to justify the use of orthopedic appliances in cleft treatment, no long-term utility has ever been established. We have included a chapter by Grayson (Chap. 21) on “Nasal Alveolar Molding” (NAM) which stresses the need to use their system to lengthen the columella. Since the shortened columella is a habilitative problem, Berkowitz believes for this reason alone NAM might have value. Unfortunately, as of this date, no long term studies have been recorded to support its use for this purpose. An unpublished

study, reported at one of the “Great Debates” that took place at the annual American Cleft Palate-Craniofacial Association meeting held in St. Louis in May 1990, “On the Thesis Presurgical Infant Orthopedics Is By and Large a Waste of Time,” Ross and Cutting argued that one of the main reasons for utilizing the procedure is for parents to believe that something important is being done to help their child and that doing something is better than doing nothing. Bardach and Berkowitz, on the other hand, argued that parents do not need to be misled and should be properly instructed to accept the need for staged treatment over a long period of time. If the only use of the appliance is to make parents feel better, those who advocate its use should say so!

A well-structured prospective clinical trial on the long-term utility of PSOT in the Netherlands by Kuipers-Jagtman and Prahll-Anderson and C. Prahll in 2003 reported:

- **Objective:** To study the effect of infant orthopedics (10+) on maxillary arch form and position of the alveolar segments.
- **Design:** Prospective two-arm randomized, controlled trial in parallel with three participating academic cleft palate centers. Treatment was assigned by means of a computerized balanced allocation method.
- **Setting:** Cleft palate centers of Amsterdam, Nijmegen, and Rotterdam, the Netherlands.
- **Patients, Participants:** Infants with complete unilateral cleft lip and palate and no other malformations.
- **Interventions:** One group (10+) wore passive maxillary plates during the first year of life; the other group (10-) did not. All other interventions were the same.
- **Main Outcome Measure(s):** The presence of contact and/or overlap (collapse) between the maxillary segments at maxillary casts made shortly after birth, at 15, 24, 48, 58, and 78 weeks. Survival experience of contact and collapse with time as well as the frequencies of different arch forms and severity of collapse were evaluated.
- **Results:** Comparable arch forms with no contact or overlap of the maxillary segments were seen at birth in both groups. With time, the frequency of collapse increased, with no significant differences between groups. No significant group differences were found with respect to the survival experience of contact and collapse or for the severity of collapse at the end of the observational period.
- **Conclusions:** Infant orthopedics does not prevent collapse and can be abandoned as a tool to improve maxillary arch form.

18.4.3 The Long-term Effect of Primary Bone Grafting

There are still divergent opinions on the benefits of primary bone grafting. The reason for the differences in part reflects the different goals of the investigators. Most of the clinical reports since have shown that buccal crossbites can still occur. Not surprisingly, the negative effect of primary bone grafting on maxillary growth is best summarized by Ross's [83] multicenter study of cases with CUCLP. It indicated that early surgical repair of the alveolus by means of infant and early childhood bone grafting or soft tissue repair of the alveolus resulted in a deficiency in the vertical growth of the anterior maxilla. When a bone graft or periosteoplasty was used, the vertical growth deficiency was found to be slightly worse and lower face height was greater, so that the vertical proportions of the resulting face were poor. Ross also reported observing a slight anteroposterior maxillary deficiency following bone graftings; however, this effect was not noted when bone grafting was delayed until 9 years of age. Surprisingly, he found that anterior maxillary vertical growth was inhibited following bone grafting, even as late as 9–13 years of age. Ross concluded that vertical growth effects can be avoided only if bone grafting is postponed until age 15 or later.

Berkowitz (unpublished data) has not found that growth problems result from bone grafting after 8–9 years of age; and the anterior vertical growth deficiency, as measured by the presence of an anterior open bite, was not commonly observed in his ongoing palatal growth study. He has, however, found anterior growth problems in cases treated with pinned appliances followed by periosteoplasty used soon after birth to retract the premaxilla in complete unilateral and bilateral cleft lip and palate.

Vertical maxillary growth leads to a more horizontal columellar plane and a reduced nasolabial angle. In individuals with unilateral clefts, this angle is almost 90° instead of the approximately 110° found in non-cleft individuals. As Bergland et al. [79] and Wolfe and Berkowitz [90] have suggested, because of the resulting midfacial growth interference, it is obviously best to delay alveolar repair at least until the mixed dentition, when secondary alveolar bone grafting offers excellent clinical benefits of closing fistulae and allowing tooth migration into the lateral incisor space.

Smahel and Mullerova [91], in a cephalometric study of males with UCLP at puberty, compared developmental changes in patients treated with primary periosteoplasty with those treated with primary bone grafting. At puberty, patients with primary bone grafts characteristically were found to have anterior

crossbites and a posterior rotation of the mandible. Deterioration of growth of the maxilla was the same in both groups, leading to the flattening of the facial profile.

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19 History of Neonatal Maxillary Orthopedics: Past to Present

ANNE MARIE KUIJPERS-JAGTMAN, BIRTE PRAHL-ANDERSEN

19.1 Introduction

More than half a century after its introduction by the Scottish prosthodontist C. Kerr McNeil [1], neonatal maxillary orthopedics still remains a controversial part of the comprehensive care for cleft lip and palate patients. The therapy, also known as presurgical or early orthopedic treatment, presurgical or infant orthopedics, early maxillary orthopedics, or more recently nasoalveolar molding, was rapidly adopted by many centers around the world, although at that time there was no scientific evidence for the claimed benefits, nor were the possible adverse effects properly investigated [2–4]. In Europe in the year 2000, about 54% of the operational centers used neonatal maxillary orthopedics [5].

Many different types of neonatal maxillary orthopedics have been described in the literature. A wide range of appliances has been designed for this purpose with pin-retained active appliances at one end of the spectrum [6, 7] and passive appliances at the other [8]. Arbitrarily, they fall into three main categories: active, semi-active, and passive appliances. Active appliances are constructed to apply a force to the maxillary segments to move them into the desired direction by using an active force delivery system like springs and screws. Additional anchorage can be obtained by pins that are driven into the maxillary bone holding the plate in position [9, 10]. Semi-active appliances are constructed by sectioning the dental cast and reorienting the maxillary segments in a more favorable position. The plate is made on the reconstructed cast and will force the palatal segments into the predetermined direction, when placed in the oral cavity. External strapping across the cleft can be part of the treatment protocol. These are the McNeil [1] and Burston [11] type of appliances. Passive appliances that are combined with extraoral strapping also fit into this

category [12]. Passive appliances are supposed to induce arch alignment during growth by grinding away material in definitive areas of the plate to ensure a proper spontaneous development of the segments. The plate is held in position by suction and adhesion only and no extraoral strapping is applied. The so-called Zurich approach as proposed by Hotz [13] and Hotz and Gnoinski [8] is the most well known representative of this kind of neonatal maxillary orthopedics.

Over the years neonatal maxillary orthopedics has given rise to emotional debates between advocates and opponents of the procedure. At the annual meeting of the American Cleft Palate-Craniofacial Association held in St. Louis in 1990, the thesis “Presurgical orthopedics is by and large a waste of time” was discussed during the so-called “Great Debate”. During the 8th International Congress on Cleft Palate and Craniofacial Anomalies in Singapore in 1997, a comparable debate took place, discussing the statement “Is presurgical infant orthopedics more a luxury than a necessity? Before and after the debates, the audiences voted equally in favor or against the statements. It was considered a major problem that hardly any sound research findings were available regarding the effects of neonatal maxillary orthopedics. And even at the 40th anniversary of one of the most talked-about critical reviews on presurgical orthopedics and primary bone grafting by Samuel Pruzansky from Chicago, what he stated still holds generally true: “The procedures advocated might be defended on the basis that continuous exploration for new and better methods is warranted and deserves support. While such research may be justified, despite the costs incurred, it is incumbent on the investigator to document his results in a scientific manner. Instead, we have been fed opinion, anecdotal pap, wishful thinking, and empirical trivia” [2].

19.2 Early History of Neonatal Maxillary Orthopedics

Before the modern school of neonatal maxillary orthopedics began, facial binding or adhesive tape strapping was used centuries ago to narrow the cleft before surgery [14]. A variation on these external bindings that were all used to reduce the cleft width was the method of Brophy [15]. He wired both ends of the cleft alveolus together in order to reduce the cleft by tightening the wire, after which lip surgery could be performed.

All procedures, which were at that time mainly done by prosthodontists or by the surgeons themselves, were based on the never proven assumption that a narrow, well-aligned cleft would be easier to repair with less undermining and less mobilization of soft tissues. A narrower cleft would also lead to less tension in the repaired lip. These claims have been repeated over and over again since McNeil started to advocate presurgical orthopedics half a century ago [1, 16, 17], although he used another argument to perform this treatment. He adopted the theory of the anatomist Scott [18, 19], who suggested that the palatal segments being detached from the downward and forward growth of the cartilaginous nasal septum, the so-called growth center, remained deficient and retruded in the face. In his opinion this resulted in the midfacial deficiency often observed in cleft lip and palate patients. By molding the palatal segments into the anatomical correct position by using a series of acrylic plates, McNeil believed that this would produce a normal maxilla while reducing the alveolar and palatal cleft at the same time.

Much confusion with respect to the effect has arisen, because the morphology of the maxilla of newborn children with clefts varies from collapsed maxillae to wide open clefts of more than 15 mm. Obviously different treatment mechanics have to be used in these cases. Burston [11, 20, 21] advocated a passive plate to start with to facilitate feeding and letting the child adapt to the plate. A following plate should be constructed by sectioning and aligning the segments of the maxilla on the plaster models before constructing the acrylic plate. These plates were used with external strapping. The plates were fitted with “wings”, the chief purpose of which was to enable the parents to insert and remove the plates and not as stated later to aid retention. McNeil and Burston claimed that soft tissues overlying the hard palate were stimulated to grow and that neonatal maxillary orthopedics could control and modify the postnatal development of the maxilla. These two practitioners have had a tremendous impact on the early treatment of children with clefts and led the way for the idea of bone grafting in conjunction with early orthopedic treatment (see also Sect. 19.4.1).

19.3 Claimed Benefits of Neonatal Maxillary Orthopedics

There has been consensus since the early 1950's that a multidisciplinary team within a center can best treat children with cleft lip and palate. The team should function as an organization with a general policy for the treatment, and each member of the team should have insight into the different aspects of treatment. The orthodontic discipline has been part of such organizations from the start, because in the majority of cases dental arch distortion and a constricted maxilla were thought to be the result of the surgical reconstruction of the lip. Presurgical orthopedic treatment or neonatal maxillary orthopedics aims at securing a good maxillary arch form in acceptable relationship with the mandible and to restore normal oral function. McNeil [1, 16, 17] and Burston [11, 20, 21] claimed that the deformation seen at birth was partly due to the original lack of tissue and a further complication of growing tissues having become mal-related consequent to cleft formation. McNeil [16] stated that the most important aspect of preoperative treatment of cleft palate infants was control and correction of the lateral segments in unilateral and bilateral conditions, and that the effect of bone stimulation plays an important role in this correction. Forces which are within the limits of biological tolerance are said to stimulate bone growth if they are applied to particular regions and in such a direction that they can be regarded as intensified normal forces.

The attainment of an end-to-end position of the alveolar processes before lip operation was the ultimate goal for neonatal orthopedics performed by all dental practitioners in the past. It has been shown that it is possible to narrow the cleft and to achieve an anatomical correct position of the maxillary segments at the time of lip surgery [8, 16, 11, 22–29]. It also improves the angulation of the palatal shelves to a more horizontal position [30–33]. However, the question remains of whether these short-term effects prior to primary surgery have any beneficial effect on overall treatment outcome in the long term.

Although neonatal maxillary orthopedics originally was instituted to restore the normal anatomy and to guide growth and development of the maxillary segments, only after the introduction of presurgical orthopedics did the orthodontic discipline try to justify early intervention for other reasons. Pahl et al. [28] summarized the arguments of the proponents of the use of infant orthopedics who state that this approach allows a more normalized pattern of deglutition, prevents twisting and dorsal position of the tongue in the cleft, improves arch form and position of the alar base, facilitates surgery, and improves outcome in general. Other alleged benefits that became en vogue are reduction of posterior cleft width, prevention of

initial collapse after surgery, prevention of cross bites, straightening of the nasal septum, facilitation of feeding, less danger of aspiration, better speech development, better nose breathing, better middle ear conditions, less extensive orthodontic treatment at later ages, and a positive psychological effect on the parents [8, 34–43]. The opponents of neonatal maxillary orthopedics stated that neonatal maxillary orthopedics is a complex and expensive therapy that is ineffective and unnecessary [2, 3, 44]. Parents are obliged to travel frequently to the treatment center and are given the burden of compliance [28]. Furthermore it is stated that neonatal maxillary orthopedics restricts maxillary development [2, 45] and influences speech negatively due to delayed surgery of the hard palate inherent to neonatal maxillary orthopedics [46, 47].

Unfortunately we have to conclude that despite half a century of research into neonatal maxillary orthopedics the results of these studies remain inconclusive, mainly due to shortcomings in the research design. Most studies were nonrandomized, retrospective in nature, have a small sample size, and lack a (randomized) control group or used historical controls. Furthermore, treatment protocols are often not properly described, and the competence of the professionals who performed the treatment is not always well documented. Also, frequently, no clear outcome measures are given and confounding variables are not taken into account.

19.4 Specific Types of Infant Orthopedics

19.4.1 Kernahan Rosenstein Procedure

An obvious problem after realignment of the maxillary segments by either neonatal orthopedics or lip surgery alone is of course to hold the realigned segments in position. For this purpose bone grafting, as a primary procedure at or around the time of lip repair, was first utilized in the late fifties in Europe [48, 49]. Great emphasis was placed on the importance of this procedure in stabilizing the realigned maxillary segments [50]. In 1977 Berkowitz [102] concluded in his state of the art report that the benefits of presurgical orthopedic therapy (PSOT) were not proven and that primary bone grafting was detrimental to midfacial growth. At that time many clinicians who previously advocated primary bone grafting abandoned the procedure because of its negative effect on maxillary and midfacial growth [51–53]. After 1975 several addition-

al centers confirmed midfacial growth inhibition and discontinued the use of primary bone grafting [54–58].

Over the years a few centers continued to use neonatal maxillary orthopedics in combination with primary bone grafting. One of these is the Children's Memorial Hospital Cleft Palate Clinic, Chicago [59]. The Rosenstein appliance is a passive plate that is inserted prior to lip surgery. Then the lip is closed and the arch segments are molded until they are in butt alignment, after which the segments are stabilized by a small subperiosteal onlay rib graft. The plate is retained for 6 to 8 weeks postgraft and the palate is usually closed at or before 12 months of age [60, 61]. In complete bilateral cleft lip and palate, the appliance covers the lateral segments, holding them in position while an extra-oral elastic band and later on the restored lip molds the premaxilla backwards.

Since 1965 Kernahan and Rosenstein have used this approach, claiming that there is a principal difference with other approaches both in sequence and technique of the procedures, and that this is critical to its success [60]. In 2003 Rosenstein and coworkers reported the long-term results in a sample of 135 patients, or about 50% of the cases that were seen in their clinic since 1965 [62]. In their paper they give an overview of the comparative studies with other centers that have been performed. These studies show that the growth in the Chicago sample was as good as for any other sample treated without primary bone grafting. Their incidence rate for orthognathic surgery was about 18%. In his multicenter study Ross [63], and later on Trotman et al. [64], found that patients who underwent primary bone grafting had on average less protrusive maxillae than the nongrafted sample, but the mandible compensated for this by downward and backward rotation, resulting in an increase of lower anterior facial height. Unfortunately the authors don't give quantitative data about the quality of the bone graft and the need for a secondary bone graft in their patients. They found in UCLP 41% of agenesis of the lateral incisor at the cleft side and in BCLP almost 50%.

It must be concluded that the value of the Rosenstein procedure of neonatal maxillary orthopedics is difficult to evaluate, as it is not a single procedure but part of a specific treatment plan that includes primary bone grafting. The long-term evaluation of the Chicago sample shows interesting results, but evaluation bias cannot be ruled out as there is an attrition rate of 50% of the original sample. More long-term studies of consecutive cases from other centers are needed to verify the reproducibility of the Chicago protocol.

19.4.2 Latham-Millard Pinned Appliance

In the Millard-Latham method of neonatal maxillary orthopedics, forces are applied using a pinned palatal appliance to manipulate mechanically the maxillary segments into close approximation, followed by alveoloperiosteoplasty and lip adhesion [65]. Latham based his treatment concept on the facial growth hypothesis of Scott [18, 19] as mentioned earlier, and he encouraged Millard to use it in complete unilateral and bilateral clefts. The aim of the procedure is 'to carry the interrupted embryonic process to normal completion' by maxillary alignment, stabilization of the alignment along with tunneling of the alveolar cleft with periosteum, and reconstruction of the nasal floor to support the alar base [66].

It was many years before the first longer-term studies were published about this procedure. Berkowitz [68] found that of a group of 32 UCLP patients at 6 years of age, 23 cases had an anterior cross bite due to the displacement of the premaxillary portion of the larger maxillary segment and the scar tissue resulting from the periosteoplasty. Comparable results were found analyzing dental casts of 63 patients with complete clefts showing a greater percentage of anterior cross bites at 6 years, but becoming less at 9 years of age compared to a lip adhesion alone group [68]. Lukash et al. [69] also found anterior and lateral cross bites after 6 years of age in most UCLP cases, but in BCLP cases arch form and occlusion as well as mid-facial growth were considered acceptable. The findings in BCLP patients are in contrast to the data from Berkowitz [68, 70], who showed in a BCLP group of 14 patients treated by Millard and Latham that all patients had midfacial retrusion at 9 years of age and in 50% of the cases the premaxilla was retruded with the anterior teeth in cross bite.

In 2004 Berkowitz and co-workers [71] published an extensive report on their findings in 30 UCLP patients and 21 BCLP patients treated with the Millard-Latham procedure at the South Florida Cleft Palate Clinic. These patients were compared to patients who were treated without neonatal maxillary orthopedics and alveoloperiosteoplasty. Both in complete UCLP and BCLP they found a higher frequency of anterior cross bite and (except at 3 and 12 years of age) also for buccal cross bite compared to the treatment alternative without neonatal orthopedics and alveoloplasty. The only other long-term follow-up study found in the literature reports more anterior open bites and posterior cross bites in 55 UCLP and BCLP patients treated with the procedure compared to a control group without the procedure [72].

In analyzing the results of the published studies, the difficult problem is that in the Latham-Millard procedure, besides neonatal maxillary orthopedics,

infant periosteoplasty is always performed, although it must be emphasized that their technique of periosteoplasty is more limited with less undermining of periosteum on the maxilla than the original one, described by Skoog [73]. However, infant periosteoplasty seems not to be widely performed in most European centers anymore, as its expectations have not been fulfilled [74–79]. Nevertheless, in the United States, the Millard-Latham method continues to attract attention despite the fact that its benefits are questionable [80]. Based on the long-term results reported by Henkel and Gundlach [72] and Berkowitz et al. [71], we conclude that there is reason to be at least suspicious of the Millard-Latham procedure or to even abandon it.

19.4.3 The Zurich Approach

In most other clinics in Europe, neonatal maxillary orthopedics started in Zurich in the mid-1950s based on the treatment principles of McNeil. After the first long-term evaluation it became clear that forced approximation of the maxillary segments was not advisable. Consequently the procedure was greatly modified to what is known now as the "Zurich approach" [8, 81]. According to Hotz and Gnoinski the primary aim of neonatal orthopedics is not to facilitate surgery or to stimulate growth as postulated by McNeil, but to take advantage of intrinsic developmental potentials. Since 1969/70, early maxillary orthopedic treatment was essential in Zurich, while surgical intervention was postponed in order to minimize subsequent growth disturbance, create optimal conditions for the maxillary segments to develop their entire growth potential, maintain or improve arch form, and control effects of surgical lip closure. The appliance used is a passive plate of compound soft and hard acrylic resin, and it is worn 24 hours a day for about 16 to 18 months, when the soft palate is closed surgically. The hard palate is closed after 5 years of age [8, 13]. During the course of treatment the lip is closed at about 6 months of age. The posterior extension of the plate that extends to the uvula must be carefully adapted to the specific anatomy of the patient. Arch alignment is achieved by grinding away the acrylic in specific areas. Figure 19.1 shows a patient who was treated according to these principles.

According to the scientific use at that time, conclusions regarding the effects of treatment were mainly based on observations: orthopedic guidance together with optimal timing of surgery has beneficial effects. Later evaluations claimed better results with two-stage palatal closure on speech; whether or not the presurgical treatment had an influence could not be demonstrated [8, 35, 82, 83]. The Brogan technique



Fig. 19.1 a–e. Patient with a complete unilateral cleft lip and palate at birth (a), after presurgical orthopedic treatment according to the Zurich approach (b), after lip closure (c), frontal view at the age of 15.8 yr (d), and palatal view at 15.8 yr after secondary bone grafting and orthodontic treatment (e)

combines the McNeil and Burston technique with the Zurich approach [24]. After alignment of the maxillary segments with a loose fitting passive appliance and extra-oral strapping, a passive plate with internally readjustments and without strapping is used.

We can conclude that the treatment principles of Dr. Hotz and Dr. Gnoinski have had a tremendous impact on cleft palate treatment, especially in Europe. Unfortunately in the last two decades not much has been published about the results of the Zurich approach, and the earlier papers have no strict research design that allows us to draw evidence-based conclusions about the effectiveness of neonatal orthopedics.

It might very well be possible that the surgical timing and sequencing are the decisive factors in the final treatment result.

19.4.4 Nasoalveolar Molding Grayson

After nearly forty years of neonatal maxillary orthopedics, the treatment started to drift away from its traditional aims and techniques with the publications from the Cleft Palate Team at New York University Medical Center in the 1990s [84, 85]. To theirs and many others' experience, it is very difficult to reach an

esthetically pleasing result in the treatment of cleft lip and palate due to the actual anatomical deformity of the nose, which includes abnormal nasal cartilage morphology, deviated nasal septum and columella, asymmetry of the alar, and a short or even absent columella, depending on the type of cleft. In addition some fibers of the orbicularis oris muscle run superiorly along the margins of the cleft towards the nasal tip, while in bilateral clefts, muscle tissue is often lacking in the prolabium, which is seated directly at the short columella. Therefore Dr. Grayson and Dr. Cutting emphasize the importance of presurgical correction of the nasal cartilage and soft tissue deformity, which can be achieved by a combination of nasal and alveolar orthopedic molding [103]. For more detailed information on this topic the reader is referred to Chapter 21 of this book.

The claimed benefits of the procedure are (a) improved long-term nasal esthetics, (b) a reduced number of nasal surgical procedures, (c) a reduced need for secondary bone grafts if gingivoperiosteoplasty is applied, (d) no larger growth disturbance than is found for other well-established procedures, and (e) savings in cost. In a series of papers that are discussed in the 2001 paper, the New York group attempted to substantiate these claims [103]. However, as for all other types of neonatal maxillary orthopedics, the research design was not adequate to show scientific evidence for these claims. In line with the DUTHCLEFT trial as described below, a two-group randomized controlled clinical trial would be the preferred design to investigate the effect of nasoalveolar molding before deciding whether to accept or abandon this therapy. Berkowitz in Chapter 17 writes strongly about the negative effects of PSOT. Unfortunately the N.Y.U. group has not provided occlusion or cephalometric studies to support the use of PSOT.

19.5 The DUTHCLEFT Study

19.5.1 Background

As noted above, neonatal maxillary orthopedics was developed and introduced on theoretical grounds, and it became part of the treatment protocol in many centers although the actual effectiveness had never been tested, nor were the possible adverse effects properly looked into [2–4, 14, 28]. We have both performed neonatal infant orthopedics according to the Zurich approach for over 20 years, but, our clinical experience has convinced us that in unilateral cleft lip and palate patients, neonatal orthopedics is at best not necessary and at its worst could have negative side effects, even if considering only the cost-effectiveness. Gradually we reached equipoise regarding whether or

not to perform neonatal infant orthopedics. Being strong supporters of evidence-based care in general, and for cleft lip and palate patients in particular, a randomized controlled clinical trial was a next logical and inevitable step to try to solve the controversy regarding neonatal orthopedics.

After obtaining funding for a first period of three years, a prospective randomized clinical trial to investigate the effect of neonatal maxillary orthopedics in children with a complete UCLP, named 'DUTHCLEFT', was begun in 1993 in three academic Cleft Palate Centers in the Netherlands: the Cleft Palate Centers of Nijmegen University Medical Center, Amsterdam Free University Hospital, and Rotterdam Erasmus University Hospital.

The following outcome variables were studied:

- A. General effects: influence on feeding, body length and weight gain, parents' satisfaction
- B. Surgical and orthodontic effects: duration of lip surgery, esthetic outcome, maxillary arch form and dimensions, maxillofacial growth
- C. Speech and language development: prelingual sound production, early speech and language development, intelligibility
- D. Cost-effectiveness: medical and nonmedical costs

All children who participate in the study are now 6 years of age. So far, results for feeding, general body growth, maxillary arch dimensions, speech and cost-effectiveness are available.

19.5.2 Experimental Design

The study was set up as a prospective two-arm randomized controlled clinical trial in three academic cleft palate centers in the Netherlands. The local Ethical Committees approved the study protocol. An extensive description of the design of the study is given by Pahl et al. [28].

The intake started in January 1993 and ended in June 1996. In total 54 babies (41 boys, 13 girls) entered the trial, 27 in each group. The patient inclusion criteria were: complete UCLP, infants born at term, both parents Caucasian and, because of the speech assessments, fluent in the Dutch language, and trial entrance within 2 weeks after birth. The exclusion criteria were: other congenital malformations (except for syndactyly), and soft tissue bands. The parents of eligible infants were informed about the trial, and written informed consent was obtained from all participants. A computerized balanced allocation method was used in order to reduce imbalance on relevant prognostic factors between the two groups. Patients were allocated based on birth weight (<3300 g or ≥3300 g) and alveolar cleft width (<8 mm, between 8 and 12 mm, or

≥ 12 mm). A computer program assigned the infant to the group with infant orthopedics (= IO⁺) or without infant orthopedics (= IO⁻).

Lip surgery was performed at 18 weeks of age according to the Millard technique. The soft palate was closed at the age of 52 weeks according to a modified Von Langenbeck procedure. Neonatal maxillary orthopedics was performed by means of passive plates, starting within 2 weeks after birth. The plate was fabricated on a plaster cast and consisted of compound soft and hard acrylic. The plate, with a small extension into the cleft nose, covered the palate and the alveolar ridges and obtruded the cleft of the hard and soft palate. The plate was worn 24 hours a day. IO⁺ children returned to the clinic every 3 weeks, to have their plates adjusted by grinding at the cleft margins to ensure proper approximation of the maxillary segments. Maxillary growth indicated the necessity for a new plate. After surgical lip closure the plate was relieved in the frontal area and re-inserted the same day. Check-up visits were now planned every 4 to 6 weeks. The plate was worn until surgical closure of the soft palate. Children in the IO⁻ group did not wear plates. They visited the clinic for an extra check-up at the age of 6 weeks as well as before and after lip repair and soft palate closure.

19.5.3 General Effects

Available information in the literature of the effects of IO on *feeding* and subsequently on *general body growth* seemed to be inadequate, and therefore feeding variables and the anthropometric variables weight and length were measured in DUTCHCLEFT. The feeding variables were measured using questionnaires that were given to the parents when the child was 0 to 2 weeks of age, and at 3, 6, 15, and 24 weeks of age. The questions concerned: amount of food, duration of the feeding, feeding frequency, feeding velocity (time measurement), feeding method, breast milk or formula, and feeding problems. We also used questionnaires to measure *parents' satisfaction* that were filled out by the mother [85]. Prahl et al. [86] report on the effects of IO on bottle feeding during the first 24 weeks of life and on weight and length during the first 14 months of life.

Parents' satisfaction was comparable for both groups. Only slight feeding problems were experienced, but there were no significant differences between babies who wore a plate and babies without a plate. Feeding velocity increased with time from 2.9–13.2 ml/min in the IO⁻ group and from 2.6–13.8 ml/min in the IO⁺ group; no significant differences were found between groups. Weight-for-age, length-for-age, and weight-for-length (z-scores) did

not differ significantly between groups, but overall the UCLP infants in both groups had significantly lower mean z-scores for weight-for-age and height-for-age than the reference during the first 14 months of life, and had lower mean values for weight-for-length after soft palate closure.

The data show that neonatal orthopedics with the aim to improve feeding and the consequent nutritional status in infants with UCLP can be abandoned. In addition, parents' satisfaction proved not to be an argument to perform neonatal orthopedics.

19.5.4 Orthodontic Effects

The purpose of this part of the DUTCHCLEFT trial was to evaluate the effect of neonatal orthopedics on maxillary arch form and arch dimensions from birth on and occlusion of the deciduous dentition in UCLP children [28, 87, 88]. Maxillary arch dimensions were evaluated by means of plaster casts of the upper jaw at the ages of 0, 15, 24, 48, and 58 weeks, and 1-1/2, 4, and 6 years. The maxillary casts were analyzed three-dimensionally by means of the Reflex Microscope (Reflex Measurement Ltd., Somerset, UK). In addition, contact or collapse of the maxillary segments, at the age of 11/2 year, were scored. Figure 19.2a,b shows a series of models from the study.

The results show that neonatal orthopedics does reduce the alveolar cleft width before lip surgery, which means that at the time of lip closure the cleft width in children treated with plates was narrower than in the other group. However, after lip closure in both groups the alveolar cleft width diminished further, and at the time of soft palate closure the maxillary arch dimensions in the two groups were comparable. The same holds true for the palatal cleft width that was also reduced during the orthopedic treatment, but after lip closure no significant differences were found between the groups anymore. Furthermore, when a plate was used the palatal vault flattened. An explanation for this finding could be that the plate keeps the tongue out of the cleft, allowing the palatal shelves to flatten in the treated group. Once the soft palate was operated, the effect of treatment disappeared and at 1-1/2 year of age the shape of the palatal vault was comparable for both groups [28].

Neonatal orthopedics also did not prevent collapse of the maxillary arch [87]. The longer-term results until 6 years of age showed no significant differences between the IO⁺ and IO⁻ groups for the 5-year-old index [89–91], which is a measure for the jaw relationship. In addition, overjet, overbite, presence of cross bites, and sagittal occlusion were comparable in the two groups [88].

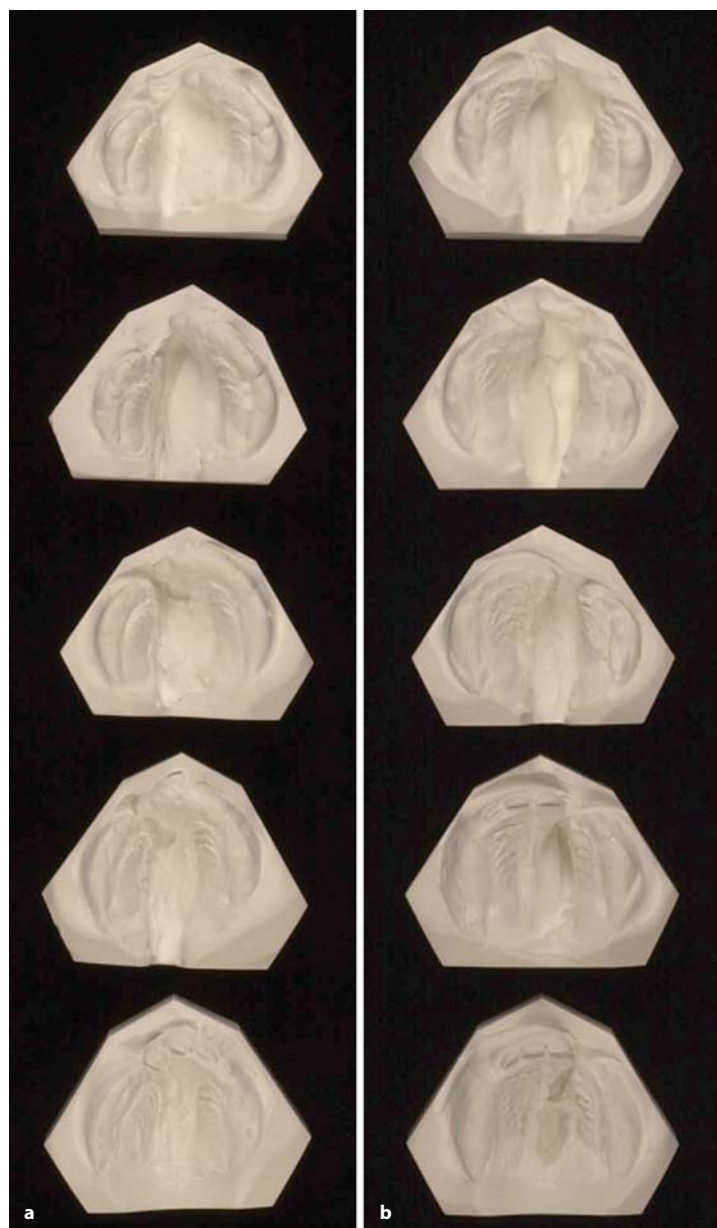


Fig. 19.2 a, b. Series of study casts of two patients from the DUTHCLEFT trial. The infant whose models are shown in **a** received no infant orthopedics; the infant whose models are shown in **b** received infant orthopedics. The five stages are (from upper to lower model): 0, 15, 24, 48, and 58 weeks of age (Courtesy Dr. Charlotte Prah, Amsterdam, The Netherlands)

We concluded that infant orthopedics has only a temporary effect on maxillary arch dimensions which does not last beyond soft palate closure. It also does not improve the jaw relationship and occlusion in the deciduous dentition until 6 years of age. Therefore from the orthodontic point of view, neonatal maxillary orthopedics could be abandoned.

19.5.5 Effect on Speech

Evaluation of speech and language development showed that at the age of 1 year the children who wore plates presented enhanced production of alveolar sounds in babbling; however, at the age of 1–1/2 year when the plate was no longer used, consonant production in babbling was comparable in the two groups [92].

Reports on the later speech and language development of the children in the DUTHCLEFT study showed a limited effect on speech [93–95]. The speech results at 2.5 years of age showed differences in intelligibility between the groups. In two different experiments, untrained listeners as well as experienced speech and language therapists gave higher intelligibility ratings to the children who formerly used plates [95]. However, data obtained by a transcription task indicated no differences in actual intelligibility [96], but compared to their non-cleft peers the children with clefts were significantly less well understood.

At 2.5 years of age, the phonological development of the IO⁺ children was normal or delayed, whereas most children who were not treated with a plate had abnormal development. Half a year later it appeared that the IO⁺ children had acquired more initial consonants than the IO⁻ group [94]. In the same age groups the IO⁺ children used longer sentences than the IO⁻ children, indicating that their grammatical development was more advanced. At the age of 6 no differences in expressive language skills between the two groups were found [93]. Hence, neonatal maxillary orthopedics did not have long-lasting effects on language development.

Regarding the claimed benefits of neonatal maxillary orthopedics on speech development in unilateral cleft lip and palate children, it can be concluded from this trial that there is a beneficial but rather limited effect until the age of 2–1/2 years. However, irrespective of neonatal maxillary orthopedics, the speech of children with clefts remains far behind that of their non-cleft peers. At this moment (2005) the speech samples at the age of 6 years have not yet been analyzed.

19.5.6 Cost-Effectiveness

Nowadays cost-effectiveness is an important issue, as costs of health care interventions are often topic of debate. Especially in cases of reimbursement decisions, cost-effectiveness estimates can be useful. Together with the results of clinical effectiveness studies, cost-effectiveness information can be used to determine whether a certain treatment should become available to patients. The main principle of cost-effectiveness analysis is to estimate the costs and the treatment outcome (= effectiveness), compared to an alternative treatment [97]. Based on this explicit comparison, the difference in costs is related to the difference in effectiveness between alternative treatments. A prospective study in a randomized clinical trial design, such

as DUTHCLEFT, in combination with an economic evaluation is a good vehicle to analyze the cost-effectiveness [98, 99].

In DUTHCLEFT a cost-effectiveness analysis was planned for all three research areas that were mentioned above (see A, B, C). For the cost-effectiveness analysis a so-called societal point of view was used, indicating that cost of treatment was based on real prices rather than fees. Besides this, a differential approach was used, aiming to calculate the difference in cost between yes or no neonatal maxillary orthopedics. A more extensive description of the cost analysis methods used in DUTHCLEFT can be found in our earlier publications [98, 100], which together with the publication by Cunningham [99] provide a good starting point to learn more about economic health evaluation.

In a preliminary report the short-term cost-effectiveness of neonatal maxillary orthopedics was based on the duration of the operation for surgical lip closure [98]. However, this effectiveness parameter has little to do with effectiveness in terms of clinical outcome, but at that time, as the trial was still running, no other effectiveness variables were available. In 2004 we published data on cost-effectiveness of neonatal maxillary orthopedics compared to no such treatment, focusing on speech outcome at 2–1/2 years of age [101]. The patients' speech was assessed by a panel of five speech therapists with experience in assessment of cleft lip and palate speech. At the time of evaluation, full data of both groups were not yet available. In each group the data of 10 children could be analyzed. The cost-effectiveness analysis required that the effects of treatment were expressed in one general effect measure. Therefore the listeners were asked to rate speech quality on a 10-point scale. A detailed description of the full perceptual evaluation is given by Konst et al. [95].

The group that was treated with neonatal orthopedics had a significantly better rating for speech (IO⁺: 3.52, SD 1.75; IO⁻: 2.18, SD 0.62). The total costs of neonatal maxillary orthopedics were of course higher in the treatment group. The resulting cost-effectiveness ratio for IO⁺ versus IO⁻ was EUR 1041.00 for 1.34 point of speech improvement. Relative to the total costs spent on the management of children with clefts, the financial investment to obtain this speech improvement is rather limited. Furthermore, the additional costs of neonatal maxillary orthopedics might be partly outweighed by the costs prevented for speech therapy in later years. This needs to be investigated further when the 6-year data of all children have been analyzed.

19.6 Conclusions

In 1991 the National Institute of Dental Research (NIDR) sought grant applications to conduct prospective and/or retrospective clinical trials evaluating treatment procedures for nonsyndromic, unilateral cleft lip and palate. The background information was that treatment sequence, timing, methods, and surgical techniques were all controversial. The long-term impact on maxillofacial growth and speech were considered to be of primary importance. This initiative reflected the lack of evidence with which treatments were carried out up to this point in time. This also holds true for neonatal infant orthopedics. Prior to the start of their randomized clinical trial of the effect of presurgical orthopedics in UCLP, we attempted to perform a meta-analysis of the existing literature, but this proved not to be possible due to the lack of randomized clinical trials, inconsistent outcomes and result reporting, missing data, small sample sizes, confounders, covariates, and publication bias. The available information on bilateral cleft lip and palate patients is even more sporadic. This was the main reason to start the DUTHCLEFT trial.

Based on the results of DUTHCLEFT so far, we conclude that neonatal maxillary orthopedics in unilateral cleft lip and palate patients as performed in this trial is not necessary for feeding, parents' satisfaction, or orthodontic reasons. Regarding speech, a positive but very limited effect was found until the age of 2-1/2 years, but the speech of the children with clefts remained far behind that of their non-cleft peers anyway. Relative to the total costs of the treatment of a UCLP patient, the financial investment to reach this effect was rather limited. However, it is questionable whether this limited effect is important enough to justify neonatal orthopedics. It should also be taken into consideration that the 6-year results for speech have not yet been analyzed.

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20 A Comparison of the Effects of the Latham–Millard POPLA Procedure with a Conservative Treatment Approach on Dental Occlusion and Facial Aesthetics in CUCLP and CBCLP

SAMUEL BERKOWITZ, MARTHA MEJIA

20.1 Dental Occlusion

One of the most widely debated areas in the treatment of cleft lip and palate involves the use of presurgical orthopedics and periosteoplasty with lip adhesion (POPLA) designed by Ralph Latham (orthodontist) and D. Ralph Millard, Jr. (plastic surgeon) [1, 2]. They contend the POPLA procedure is superior to a conservative nonpresurgical orthopedic treatment (non-POPLA) for producing more aesthetically appealing lip/nose surgery, while still allowing for good midfacial growth and dental occlusion in complete unilateral cleft lip and palate (CUCLP) and complete bilateral cleft lip and palate (CBCLP) patients. This study compares the dental occlusion of POPLA with conservative, nonpresurgical orthopedic treatment (non-POPLA), which Millard had been using between 1960 and 1980 [3]. POPLA proponents and others, who may not resort to a lip adhesion but who still use the same presurgical orthopedic procedure designed by Latham in CBCLP patients for forcefully retracting the protruding premaxilla, favor the attainment of improved facial aesthetics and palatal arch alignment soon after birth with or without periosteoplasty. They speculate that the early aesthetic benefits will remain as the face grows and develops [4–7].

In CUCLP and CBCLP presurgical orthopedics is usually followed by periosteoplasty in the expectation that the resulting bone bridging created will avert the need for secondary alveolar bone grafts at a later date.

It is not a simple or a lightly assumed task to offer a brief challenging the rationale for presurgical orthopedics and periosteoplasty for infants with complete unilateral or complete bilateral cleft lip and palate. Advocates of the POPLA concept, or a variant of it, are few but well respected. Their proposed goal of making things right and whole as early as possible seems sensible and has great emotional appeal.

This first of a two-part serial retrospective dental occlusal and facial study covers more than 40 years of

recording with serial dental casts and lateral cephaloradiographs the sequential influences of both the POPLA and conservative non-POPLA procedures on palatal development, and the anterior and buccal dental occlusion. Part One of this study uses serial dental casts to determine the extent of anterior and buccal crossbites. Detailed dental occlusal analyses of these records test the efficacy of the POPLA procedure compared to the non-POPLA treatment employed by Millard and Berkowitz from 1960 to 1980 prior to the POPLA treatment years.

Part Two of this study will analyze facial changes using serial lateral cephaloradiographs.

20.1.1 Method and Materials

The complete unilateral and bilateral clefts of the lip and palate cases, treated by either presurgical orthopedics (POPLA) or the nonpresurgical orthopedics (non-POPLA) procedure, were chosen from the files of the longitudinal facial and palatal growth studies of the Miami Craniofacial Anomalies Foundation of patients from the South Florida Cleft Palate Clinic, the University of Miami School of Medicine. D. Ralph Millard, Jr. performed lip, nose, and palatal surgery in both test samples. Secondary alveolar bone grafts and maxillary and/or mandibular osteotomies and maxillary distraction osteogenesis were performed by S.A. Wolfe. Samuel Berkowitz documented growth changes with dental casts, lateral cephaloradiographs, panorex, and photographs, and performed all treatment orthodontics other than presurgical orthopedics [4, 6]. Berkowitz treated a number of the children in the POPLA group who had extensive anterior crossbites starting when they were about 9 years of age so that beginning at this age there is a reduction in the frequency of the cases with anterior crossbites.

20.1.1.1 POPLA – Presurgical Orthopedics with Lip Adhesion [6] (Fig. 20.1 a, b)

Ralph Latham supervised the plastic surgery residents in the manipulation of the palatally pinned presurgical orthopedic appliance. Another orthodontist later performed the same relatively simple procedure. Because of the training and close supervision involved in the treatments given, there was relatively little variation in this procedure over the years covered by the POPLA data. In CBCLP, the appliance mechanically expands the lateral palatal segments, allowing for the retraction of the protruding premaxilla

into position within the alveolar arch (Figs. 20.2–20.6). In CUCLP cases, the mechanical forces bring the premaxillary portion of the larger segment medio-palatally, and in most instances the lesser segment is carried forward 2–3 mm to make contact with each other (Fig. 20.5 a, b). Afterward, the floor of the nose is surgically closed and a periosteoplasty is performed to permit the migration of alveolar osteoblasts to bridge the alveolar gap space. After the premaxillary retraction, a lip adhesion is performed followed 6–8 months later by definitive lip surgery with “forked” flaps.

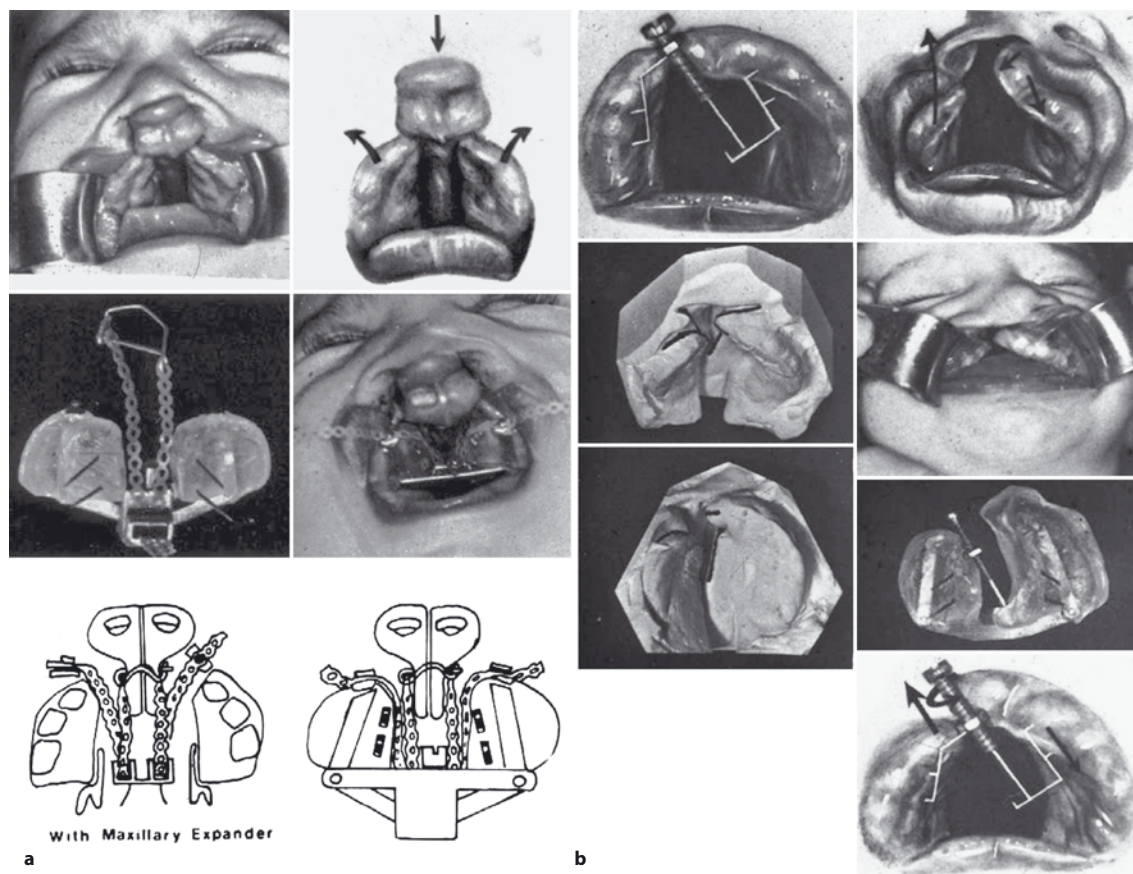


Fig. 20.1. a CBCLP. Latham's presurgical orthopedic appliance. The elastic chain creates the activating forces to retract the premaxilla while expanding the palatal segments. The posterior segment is pinned to the palate for approximately 2 weeks. The premaxillary pins, which are pulled by the elastic chain, are positioned anterior to the premaxillary vomerine suture.

b CUCLP. Latham's presurgical orthopedic appliance. The screw knob controls the movement of the pinned appliance. The premaxilla is bodily rotated medio-palatally while the cleft lesser segment is moved anteriorly approximately 2–3 mm to make contact with the smaller cleft segment

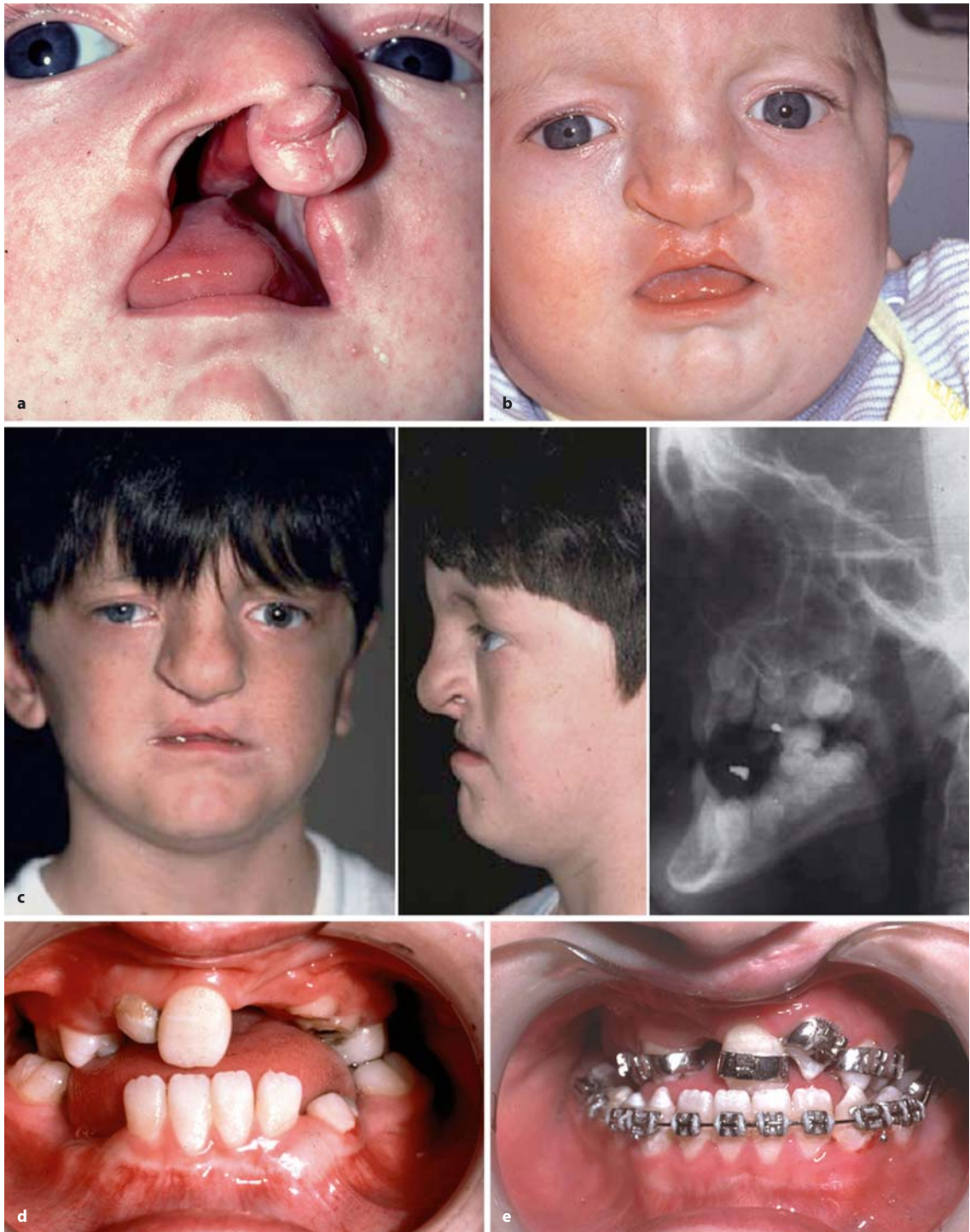


Fig. 20.2a-j. **a** Newborn. A small asymmetric protruding premaxilla. **b** After premaxillary retrusion. Lip adhesion followed by definitive lip surgery, and gingivoperiosteoplasty (POPLA). **c** Midfacial deficiency due to retruded premaxilla. **d** Retruded premaxilla showing some missing incisor teeth. **e** Orthodontic appliances placed to correct anterior crossbite and missing

incisor spaces. Protraction facial mask with Class III mechanics was unsatisfactory in correcting this problem. The use of protraction facial mask was not successful in reducing midfacial deficiency due to upper and lower facial growth. This necessitated the use of maxillary distraction osteogenesis

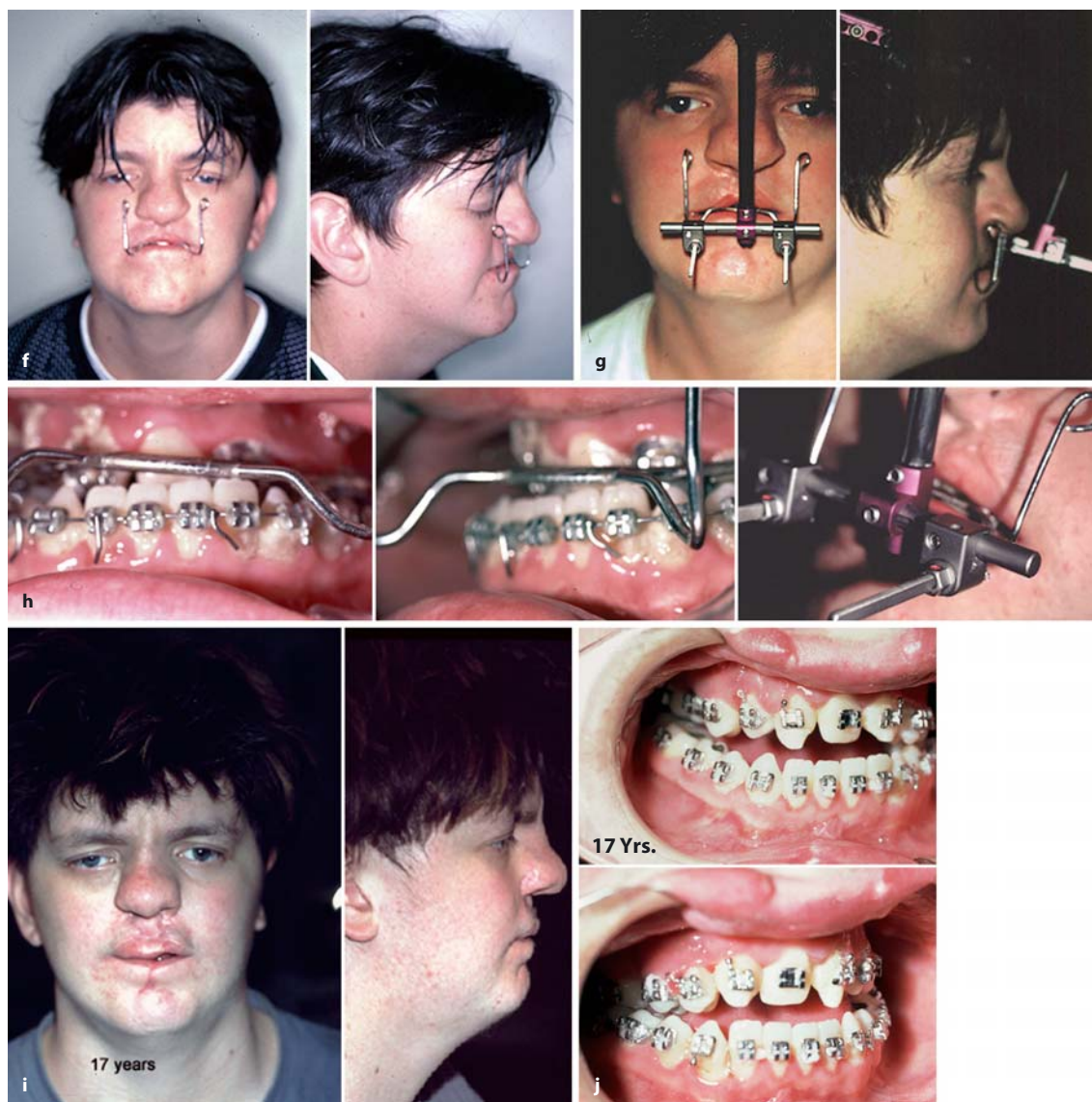


Fig. 20.2 a–j. (continued) **f, h** Osteogenic appliance. **g** After maxillary advancement. **i, j** Following distraction osteogenesis to advance the maxillary denture. **i** After protraction facial mask – no changing. **j** Distraction osteogenesis to tip occlusion – now hypernasality is present

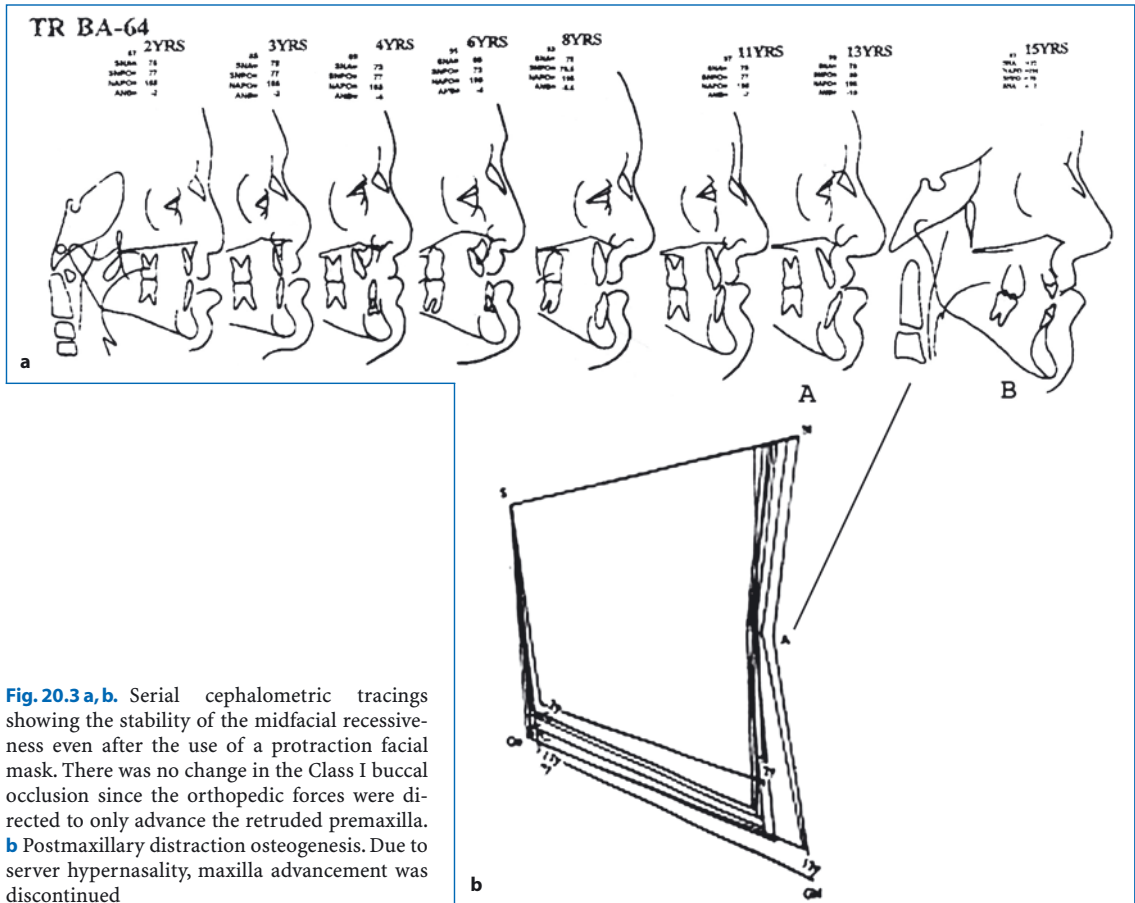


Fig. 20.3 a, b. Serial cephalometric tracings showing the stability of the midfacial recessiveness even after the use of a protraction facial mask. There was no change in the Class I buccal occlusion since the orthopedic forces were directed to only advance the retruded premaxilla. **b** Postmaxillary distraction osteogenesis. Due to severe hypernasality, maxilla advancement was discontinued

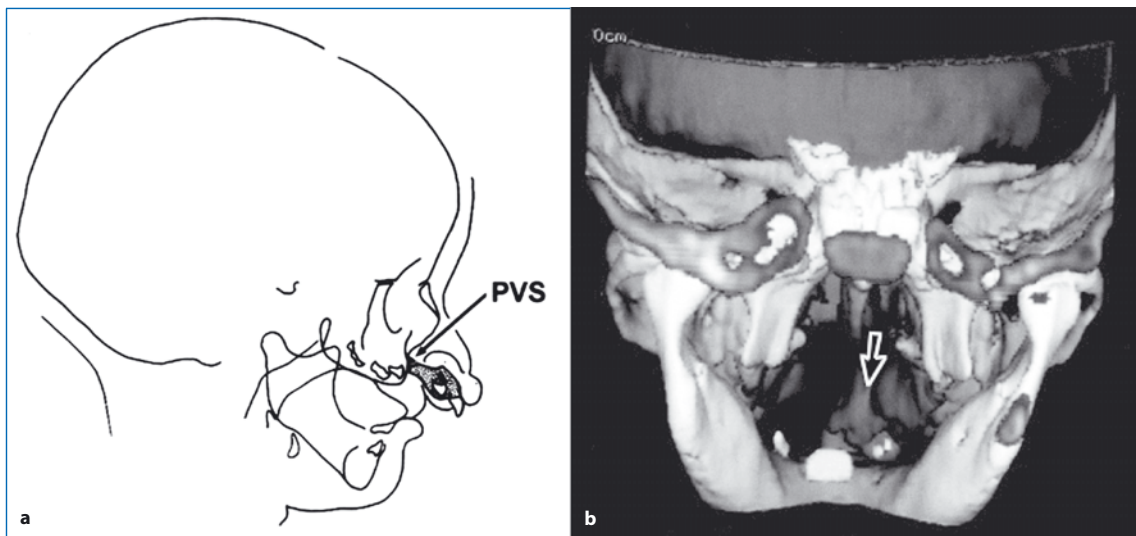


Fig. 20.4. a Lateral cephalometric tracing of a newborn with a complete bilateral cleft lip and plate shows the location of the premaxillary vomerine suture (PVS) posterior to the protruding premaxilla. **b** Frontal computed tomography scan of a patient with complete bilateral cleft lip and palate who was treat-

ed with the presurgical orthopedics, gingivoperiosteoplasty, and lip adhesion protocol. Only the left palatal segment is fused to the premaxilla with a bone bridge. Note the premaxilla "telescoping" at the premaxillary vomerine suture (arrow), the junction with the nasal septum

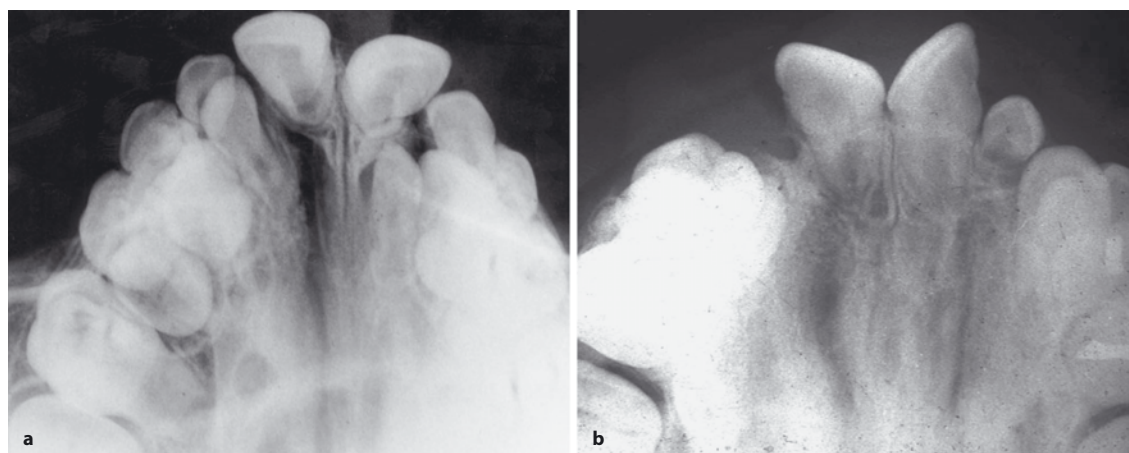


Fig. 20.5. **a** Palatal radiograph of a patient with complete bilateral cleft lip and palate who was conservatively treated at birth with a head bonnet with an elastic strip over the protruding premaxilla. The premaxillary vomerine suture was still open

years later. **b** Palatal radiograph after premaxillary retraction with a Latham appliance shows a synostosis of the premaxillary vomerine suture

20.1.1.2 Non-POPLA Conservative Treatment of CBCLP and CUCLP

The protruding premaxilla in CBCLP is ventroflexed by the forces generated by a head bonnet with elastic strip positioned across the prolabium, followed by lip adhesion surgery. No attempt is made to bodily retract the premaxilla and place it within the alveolar arch. Palatal cleft closure using a von Langenbeck procedure with a modified vomer flap is performed between 18 and 36 months, depending on the size of the cleft space. In non-POPLA as well as POPLA cases palatal expansion is sometimes used at 5–6 years of age to correct the buccal crossbite in both groups.

Orthodontics between 8 and 10 years of age on POPLA and non-POPLA patients is used to align the anterior teeth. In non-POPLA cases it is performed prior to bilateral or unilateral secondary alveolar bone-grafting. In POPLA cases the anterior crossbite correction with full orthodontic appliances and a protraction facial mask is initialized at 8–9 years when one or both of the permanent central incisors are erupted. An attempt at this age is made to correct the posterior occlusion. None of the test groups were in the Class III buccal occlusion at this age. Standard orthodontics follows to align all the permanent teeth. In CUCLP and CBCLP, lip adhesion is mostly performed at 3 months in both series followed by definitive lip surgery at 6–8 months of age.

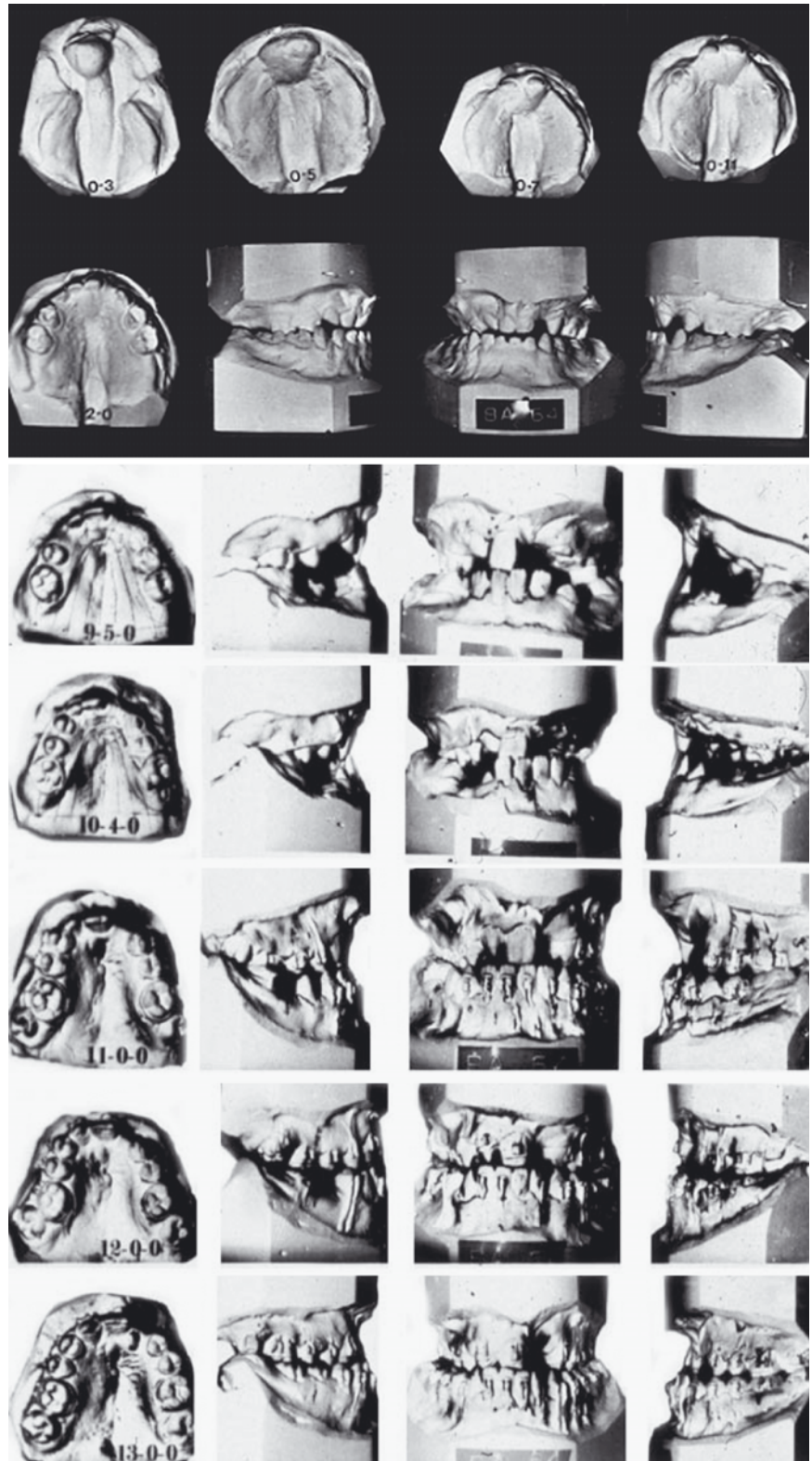
20.2 Discussion

Millard and Berkowitz have been associated since 1961 with the South Florida Cleft Palate Clinic, where presurgical orthopedic treatment (PSOT) was not being used. Millard has always been a strong supporter of the importance of serially documenting treatment outcomes with lateral cephalometric radiographs, dental casts, panorex, and facial/intraoral photographs. To that end they have worked diligently.

After 20 years of performing nonpresurgical orthopedics (non-PSO) prior to the utilization of the POPLA treatment procedure, Millard published an operational plan to investigate and record facial-palatal changes when utilizing the Latham procedure [8]. Today, Berkowitz has assembled an extensive collection of serial records that have always been available for review by any interested party. Kai Henkel [9], a visiting professor of plastic surgery from Rostock, Germany, after reviewing the POPLA-treatment serial case records, published a review stating that the procedure resulted in unsatisfactory facial aesthetics and dental function.

After 40 years of recording facial, palatal, and dental growth changes in POPLA and non-POPLA cases, the authors of this brief believe that criticism is in order for a clinical procedure whose outcome results have now been adequately reviewed using serial objective records.

Fig. 20.6. Presurgical orthopedics, periosteoplasty, and lip adhesion (POPLA). The premaxilla was retruded and positioned within the arch. Followed by periosteoplasty, the palatal cleft was closed at 2 1/2 years using a von Langenbeck procedure with a modified Vomer Flap. 9–13 years: The use of a protraction facial mask to advance the premaxilla was unsuccessful. This was followed by mid-facial distraction osteogenesis



It is unfortunate that a controversy still exists as to the utility of POPLA or similar treatment with or without lip adhesion 20+ years after its introduction, and after the critiques to which it has been subjected [3]. First, Georgiade and Latham [10] and Latham [11, 12, 13] have failed to publish any outcome studies. More recently, Millard and Latham have published a limited outcome report using Berkowitz's palatal cast records [16]. Millard and Latham's coauthors Huifen, Spiro, and Morovic performed linear measurements of the changing palatal size rather than analyze the relative growth of the opposing jaws by reviewing the dental occlusion.

Dufresne and So, in their chapter supporting the presurgical orthopedic procedure for CBCLP and CUCLP patients, referred only to Millard's original introductory statements of the POPLA procedure in *Cleft Craft* (Vol. III) [1], and listed no other supporting references, yet advocates its use [5]. Cutting and Grayson [14, 15] limited their PSOT report to the effect of periosteoplasty in successfully producing bone-bridging of the alveolar cleft; yet no mention was made of its effect on the dental occlusion.

Berkowitz [3, 16], in a preliminary report compares Millard POPLA with Millard-Berkowitz's conservatively (non-POPLA) treated CBCLP outcomes. That comparative outcome study confirmed the negative effects of POPLA on facial aesthetics and dental occlusion. The study presented herein expands on that report.

In 1996 Berkowitz and Latham were asked by the American Cleft Palate Craniofacial Association program committee to debate the utility of this procedure at their annual meeting in San Diego, California. At that meeting, Berkowitz and LaRossa (plastic surgeon) presented long-term case reports that were critical of the procedure, but no supporting case studies were forthcoming from Latham and Morales (plastic surgeon).

Many clinicians who advocate the use of Latham's or any other presurgical orthopedics (PSO) state that one of the appliance's benefits is to prevent "collapse" of the lateral palatal segments. The frequent use of the word "collapse" to describe palatal arch relationships after neonatal lip surgery is unfortunate and needs to be better understood. The term is misleading for it conjures up an unwarranted sense of foreboding.

The word "collapse" was introduced into the cleft-treatment lexicon in the 1960s by some surgeons and orthodontists to describe the palate's physical state after uniting of the lip and causing medial palatal movement of overexpanded palatal segments. They evidently did not realize that in complete lip and palatal clefts, the resulting medial palatal movement (molding) is beneficial because it reduces palatal cleft size and corrects the overexpanded palatal segments rela-

tionship. Serial studies have shown that overlapping palatal segments in the deciduous and mixed dentition is of no clinical importance.

The word "collapse" implies that this condition is bad and should be prevented. However, after years of analyzing serial dental casts, many orthodontists have concluded that establishing lip muscle continuity leads to GOOD geometric palatal changes no matter their temporary neonatal geometric relationship. Overlapped segments do not impede normal palatal growth, and in most cases such overlapped segments can easily be properly realigned using relatively simple orthodontics. Cleft segments in posterior cross-bite at an early age are not indicative of future palatal maldevelopment. Since the width of the cleft space influences the type and timing of surgical palatal closure, one would prefer to have a small cleft space prior to palatal surgery to reduce the possibility of creating growth-inhibiting scarring while producing a normal palatal vault space.

In contrast to conservative non-POPLA cases, POPLA-treated patients require extensive and costly orthodontic treatment to correct the anterior cross-bites, regain lateral incisor spaces, and achieve upper to lower anterior arch congruency. Often the degree of facial and palatal distortion is so extensive in POPLA cases that additional surgical intervention is necessary at a later age. Some parents have reported their children's experiencing psychosocial problems due to the lack of peer acceptance of the concave facial profile.

POPLA in CBCLP (Table 20.1): The bodily-retracted protruding premaxilla is retracted and placed in excellent alignment within the alveolar segments (Fig. 20.1 a, b). Since no lateral flexion of the nasal septum is seen in cat scans, it was concluded that the premaxilla is "telescoped" posteriorly at the premaxillary vomerine suture (PVS) (Fig. 20.4). The bending at the PVS is also seen in POPLA-treated CUCLP (Fig. 20.5). The PVS is not observed in follow-up palatal POPLA radiographs (Fig. 20.5).

The diagram (Fig. 20.7) is designed to show the error of palatal segmental movements in POPLA CUCLP cases. Our 3D analysis of the CUCLP palatal arch changes demonstrate that the premaxillary portion of the noncleft segment is brought medio-palatally while the smaller cleft segment is advanced, resulting in the loss of the lateral incisor space. This explains why an anterior dental crossbite is most likely to result.

Millard [1] has written that premaxillary retraction and periosteoplasty might have a negative effect on palatal growth, but he nevertheless believes this trade-off is acceptable to obtain early aesthetics, to close the floor of the nose, and to avoid the need for secondary alveolar bone-grafting.

Table 20.1. Number of cases at each age level in presurgical and nonpresurgical orthopedics treatment groups

	Approximate age of participant				
	3 years	6 years	9 years	12 years	Total sample
Unilateral cleft					
POPLA treatment	30	43	34	18	125
Non-POPLA treatment	51	54	46	33	184
Bilateral cleft					
POPLA treatment	21	20	15	9	64
Non-POPLA treatment	49	49	40	35	173

POPLA; presurgical orthopedics gingivoperiosteoplasty and lip adhesion.

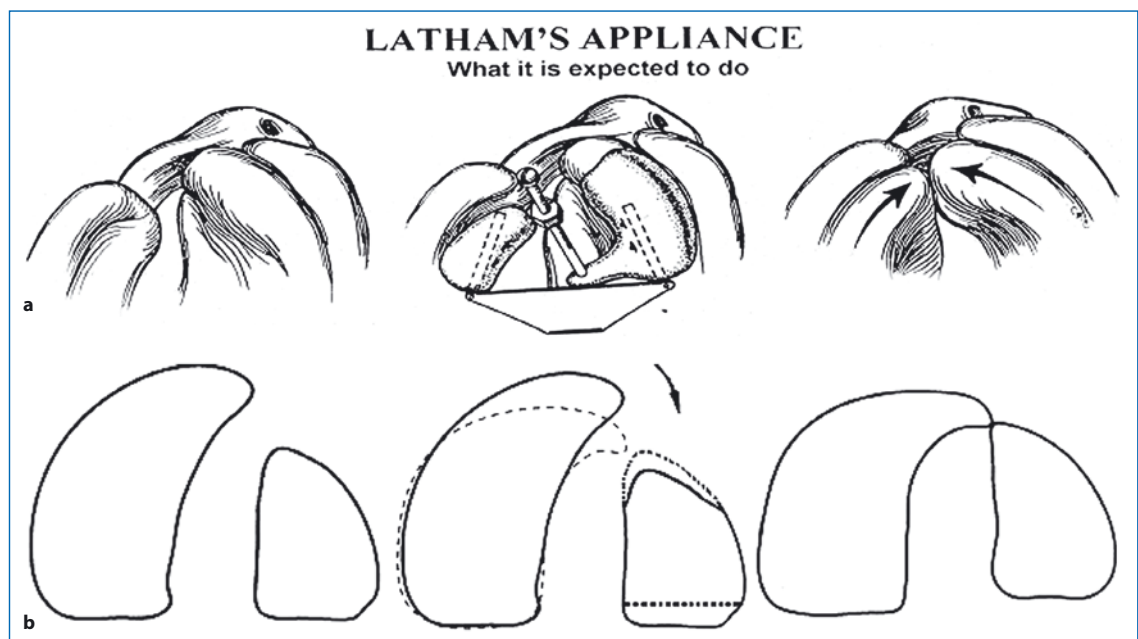


Fig. 20.7. a According to Latham, the mode of action of the appliance used in complete unilateral cleft lip and palate is the medial movement of both segments toward each other [5].
b None of our unilateral cleft cases, when activated with the

Latham appliance, is shown below. The premaxillary portion of the noncleft segment is bent medio-palatally, while the lesser-cleft segment is carried slightly forward, into contact

The desire of some surgeons to establish a child's well-balanced, aesthetically pleasing lips and nose soon after birth is understandable. However, extensive facial growth studies make clear that this should not be the top priority – that is, at the cost of good midfacial growth, dental occlusion, and speech at adolescence [3, 17–27]. All of these goals should be possible without sacrificing.

This comparative study contradicts the belief that well-designed lip/nose surgery with Latham's PSO performed on newborns to achieve early facial aes-

thetics while damaging palatal growth sites will lead to excellent adult facial aesthetics and dental function. But, should the face fail to grow well, the supporters of POPLA suggest that it can be easily corrected without far-reaching consequences such as developing poor self-esteem as a result of having a retrusive mid-face. This condition does not lend itself to easy correction by midfacial surgery alone. Extensive postsurgical psychosocial therapy may be necessary in children with inadequately formed faces [28].

In POPLA cases a buccal crossbite mostly involved the deciduous cuspid. The percentage of anterior crossbite cases increases with time (Fig. 20.29a,b). In some POPLA cases when the premaxilla is not placed precisely within the arch, less bone bridging and fewer anterior crossbites occurs. None of the cases show a Class III buccal (posterior) occlusion. The anterior dental crossbite in POPLA CBCLP and CUCLP is always due to the manipulated premaxilla's retrusive position, which is never self-correcting or could be orthodontically corrected in the deciduous or, in most cases, even in the permanent dentition. In vertically growing faces the anterior crossbite is orthodontically correctable in most cases. When the mandible grows forward, as it does in most faces, the anterior crossbite worsens requiring often a maxillary advancement surgery to create a proper dental overjet.

A typical CBCLP case of serial cephalometric tracings (Figs. 20.20, 20.28) shows an early retrusive midface in the deciduous dentition, creating an anterior crossbite with a concave facial profile that worsens with time. An extensive cephalometric report will be forthcoming in Part Two of this overall study. In these CBCLP and CUCLP cases the upper and lower face gradually grows forward but the palatal length between the first molars and incisors remains constant (Figs. 20.30–20.32). In most cases LeFort I surgery or maxillary advancement with distraction osteogenesis is necessary, as it was in this CBCLP case. In the POPLA series, maxillary distraction osteogenesis is seldom utilized in CUCLP. This surgery results in immediate hypernasality due to an increase in pharyngeal depth leading to velopharyngeal incompetency (VPI). The hypernasality slowly diminishes in some cases within 1 year, but is not completely absent in all of these cases.

In CBCLP 57% of POPLA patients, but only 18% of the non-POPLA treated cases, exhibit an anterior crossbite at 6 years of age (Figs. 20.29–20.31, Table 20.1). Posterior (buccal) crossbites are not always related to POPLA (Fig. 20.29). The treatability of buccal crossbites is also influenced by the extent of scarring created by the surgical cleft closure procedure. If a buccal crossbite is present, some may be easily corrected by 6–9 years of age in both test groups. As in CBCLP, the CUCLP shows slight anterior palatal growth. Most palatal growth occurs posteriorly to accommodate the developing molars (Figs. 20.30–20.32).

20.3 POPLA CUCLP

(Figs. 20.5, 20.8–20.15, 20.23–Table 20.1)

Early anterior dental crossbite in the deciduous dentition is associated with the loss of the lateral incisor space brought on by PSOT. This results from the medio-palatal positioning of the premaxillary portion of the larger noncleft segment, which is brought into contact with the forward, positioned lesser-cleft segment. The following periosteoplasty creates extensive bone bridging in over 80% of the cases.

POPLA patients have greater transverse posterior arch width earlier than what is observed in non-POPLA cases. This is due to the palatal appliance preventing the neonatal overexpanded lateral segments from molding together and closing off most of the palatal cleft space. There is no clinical advantage for maintaining this increased palatal width at this early age since POPLA and non-POPLA cases will eventually attain ideal buccal occlusion after relatively simple orthodontics.

An anterior dental crossbite with some degree of midfacial retrusion occurs in 60% of the CUCLP POPLA cases by 6 years of age (Fig. 20.29a, Table 20.1), while only 17% of non-POPLA-treated cases experience an anterior crossbite (Fig. 20.29b, Table 20.1). In some POPLA cases that did not result in an anterior crossbite, the lateral segments are not in contact at the time of periosteoplasty. Slight or no bone-bridging with a good lateral incisor space is associated with good incisor overjet with no midfacial retrusion. The loss of the lateral incisor space can be anticipated since alveolar bone is lacking in all cleft alveoli at birth, and for any bone bridging to occur the alveolar segments need to be placed in contact. In these cases the posterior palate is unaffected by PSO and is usually in a Class I or Class II occlusion (never in Class III) at 6 years of age.

Anterior dental crossbite correction in POPLA-treated CUCLP and CBCLP cases requires the advancement of the premaxilla in CBCLP or the premaxillary part of the noncleft segment in CUCLP. In CBCLP POPLA cases the premaxillary segment, as a result of the early palatal manipulation and periosteoplasty of the closed-over cleft lateral incisor spaces, cannot accommodate the impacted lateral incisor, if present (Fig. 20.9). This space cannot be regained in the mixed dentition and only after extensive orthodontics in the permanent dentition in 50% of the cases (Figs. 20.16–20.22, 20.26–20.28).



Fig. 20.8. **a** After appliance activations: lip and nasal distortions reduces when the palatal segments are moved together. **b** Alveolar segments in contact periosteoplasty is then performed. **c** Lip adhesion to establish muscle continuity and mold the overexpanded palatal segments in a preparation for a definitive lip surgery at 6 months of age. There are instances when

defective lip and nose surgery can be performed without first performing a lip adhesion. **f, g** Note that the upper lip is slightly retruded when compared to the lower lip. This is associated with the retraction of the premaxillary portion of the noncleft palatal segment



Fig. 20.9. **a** Distorted nostrils and lip due to aberrant muscle pull. **b** Lip adhesion reduces nasal and lip distortion. **c** Rotation advancement, the definitive lip closure. **d–g** Facial photographs at different ages. Note: The vertical facial growth pattern creates

an elongated antero-facial height. As a result the midface does not appear to be retrusive. A facial protraction mask was utilized with intraoral palatal expander to advance the maxillary incisors



Fig. 20.9. (continued)



Fig. 20.10 a–i.



Fig. 20.10 a-i. (continued) **a, b** The retracted premaxillary portion of the nonleft segment was brought mediolaterally, blocking out the right lateral incisor space. The superior torque to both palatal segments created an anterior crossbite by the forces generated by the Latham appliance. **c, d** The crossbite became more noticeable when the deciduous teeth were lost and the permanent incisors began to erupt. **e** Vertical elastics were used to extrude the upper anterior teeth. **f, g** The displaced anterior teeth were brought into alignment. The maxillary central incisors were advanced, opening the lateral incisor space. **h** The

thin bony alveolar bridge created by the periosteoplasty permitted the right and the left palatal segments to be moved later and the lateral incisor space opened. **i** A lower incisor was extracted and the incisors were retracted in order to create an overbite and overjet. An anterior open bite resulted due to the original torque of the palatal segments. The correction of the alignment of the upper incisor teeth was not stable even though an upper retainer was worn. Comment: Occlusal stability is dependent on the position of the basal bone

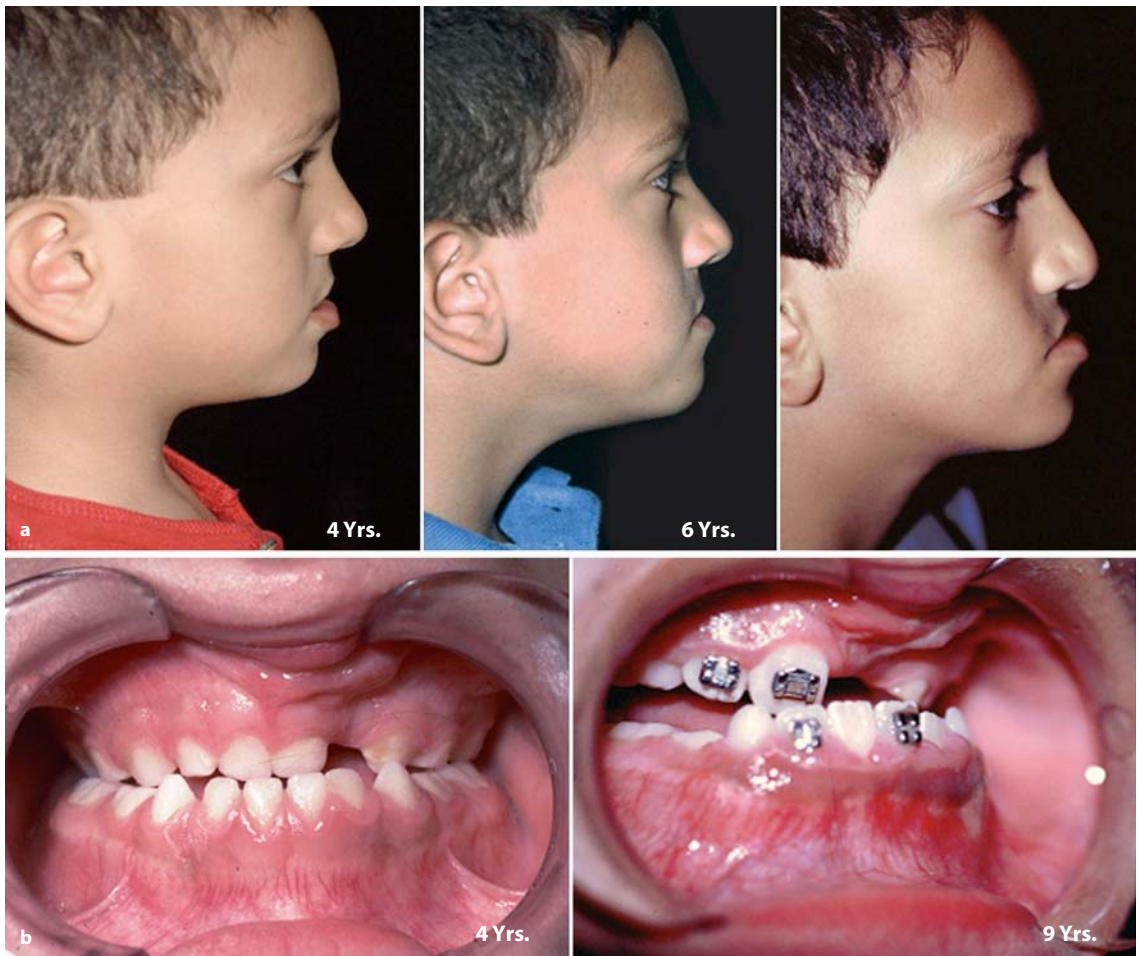


Fig. 20.11. a Facial changes. These show recessive midfaces at 4, 6, and 9 years of age. **b** Intraoral occlusal photographs showing anterior dental crossbites at 4 and 9 years of age

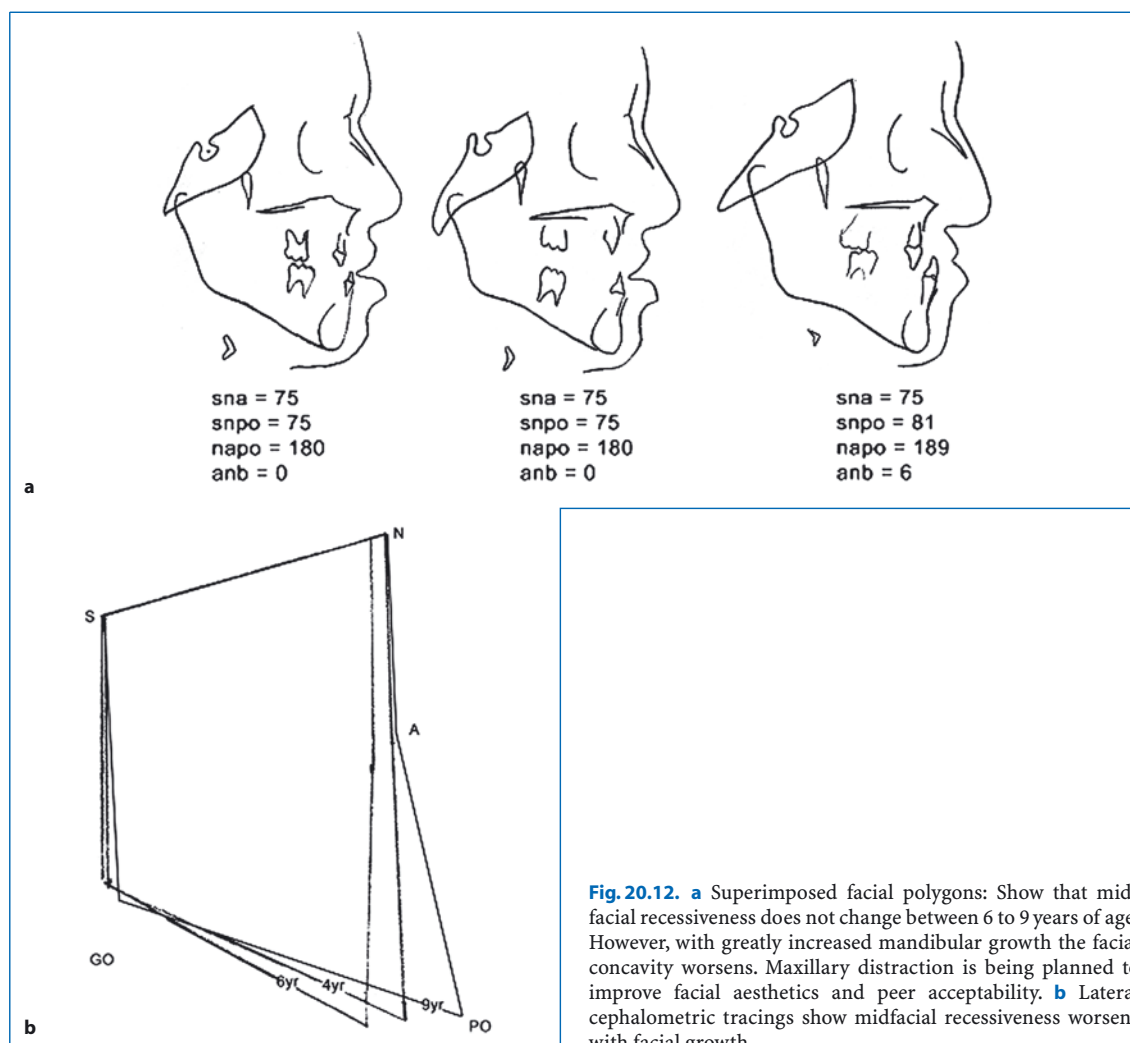


Fig. 20.12. a Superimposed facial polygons: Show that midfacial recessiveness does not change between 6 to 9 years of age. However, with greatly increased mandibular growth the facial concavity worsens. Maxillary distraction is being planned to improve facial aesthetics and peer acceptability. **b** Lateral cephalometric tracings show midfacial recessiveness worsens with facial growth

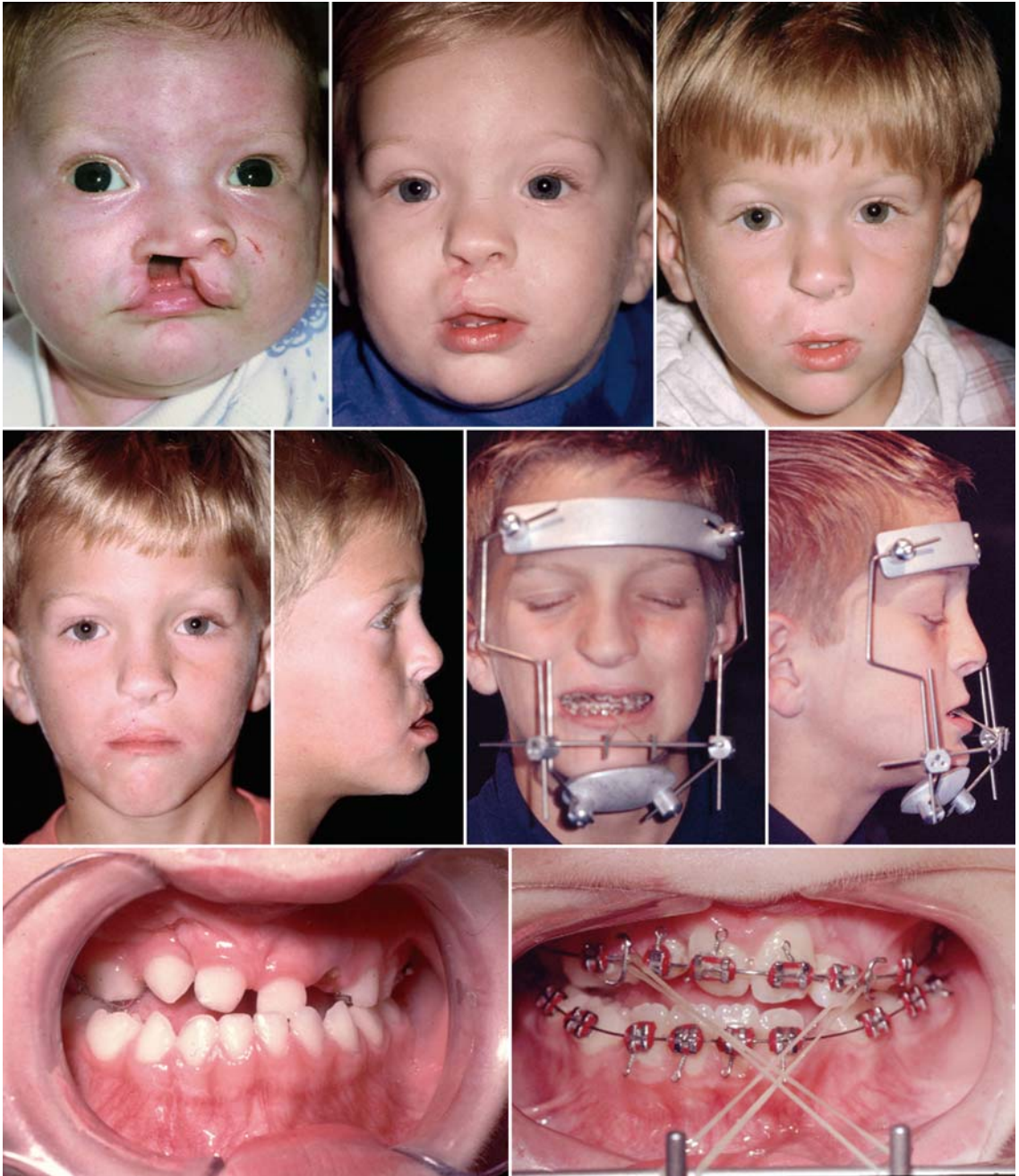


Fig. 20.13. Case BR 74. Complete unilateral cleft lip and palate. Severe anterior crossbite with crowding of incisor teeth and partial closure of the lateral incisor space. Treatment plan: uncrowd and advance incisor teeth using a protraction facial mask

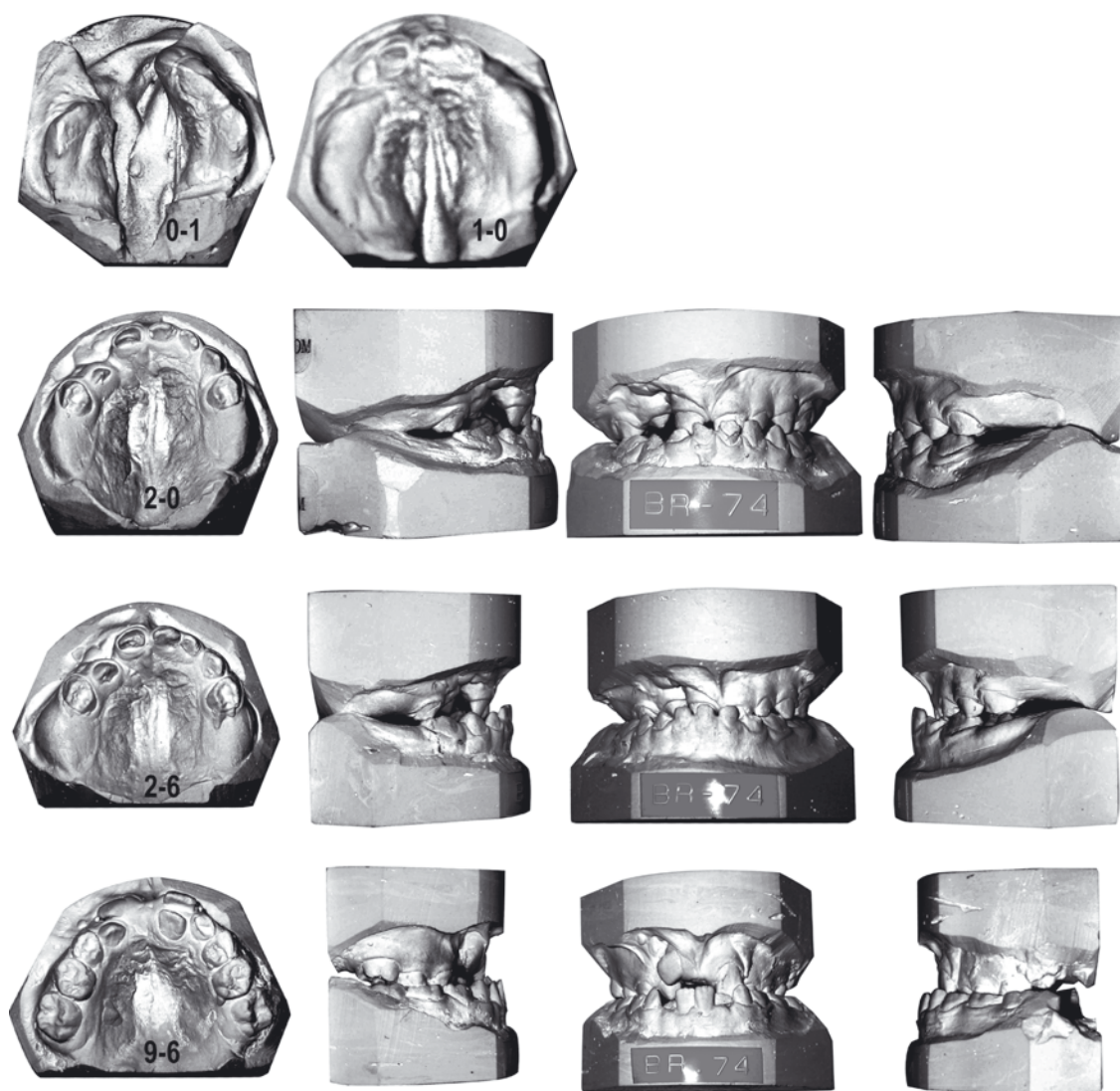


Fig. 20.14. Case BR 74. Latham-Millard Presurgical Orthopedics Periosteoplasty with Lip Adhesion (POPLA) procedure. Serial casts reflect the creation of a severe anterior crossbite and anterior dental crowding. 0-1: At 1 year after the POPLA treatment, the premaxilla portion of the larger segment was brought palatally by the presurgical orthopedics. 2-0: 2 years of age. Severe anterior crossbite with partial closure of the right lateral incisor spaces. Palatal cleft still open. 2-6: 2 years and 6 months of age. Palatal cleft space closed with a von Langenbeck and

modified vomer flap. Severe anterior crossbite with closure of the right lateral incisor space. No posterior crossbite. Comments: One can predict that the anterior crowding will become more severe with the eruption of the permanent incisors. The advancement of the anterior teeth is dependent upon premaxillary advancement, which is hindered by the bonny bridge created by the periosteoplasty. A protraction facial mask will be necessary to advance the premaxilla. (see facial photographs)

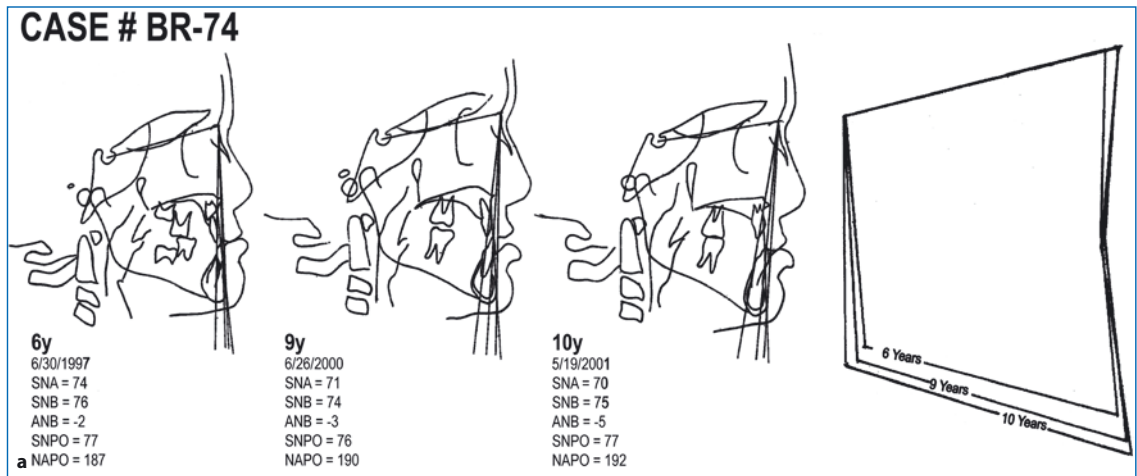


Fig. 20.15. a Case BR 74. Complete unilateral cleft lip and palate. Severe anterior crossbite with crowding of the maxillary incisor teeth as a result of retracing the premaxillary portion of the larger segment. This created a concave facial profile. (Cephalometric Analysis). **b** Case BR 74. Periapical radiograph. 7 1/2 years of age. Severe dental crowding of the maxillary anterior teeth

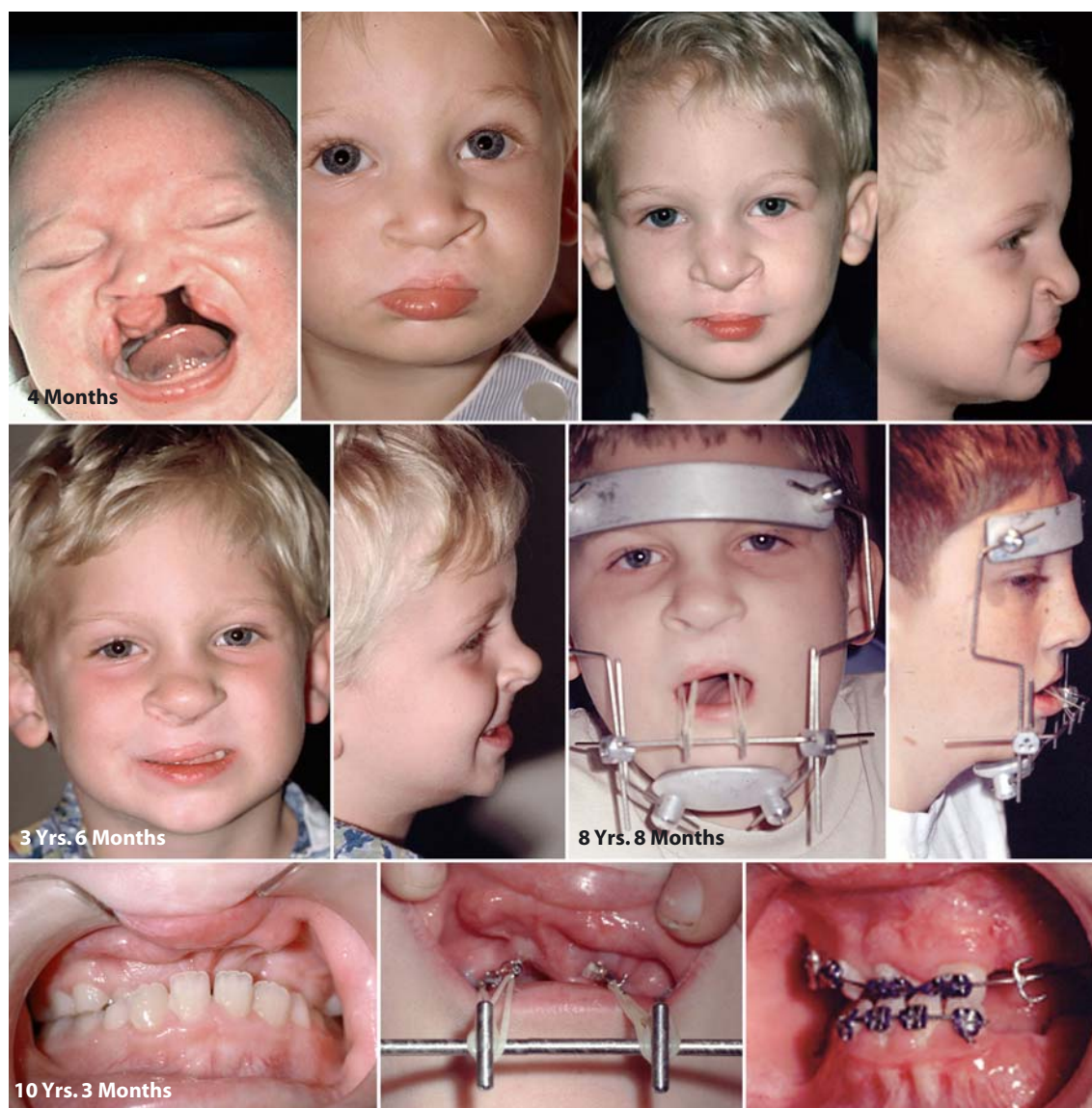


Fig. 20.16. Case BT 80. Complete bilateral cleft lip and palate. The protruding premaxilla was set back and aligned within the arch using the POPLA procedure. By 4 years of age a severe anterior crossbite resulted. At 10 years of age the teeth in the unerupted severely ventroflexed premaxilla still failed to erupt.

This necessitated the placement of a gold chain on the imbedded central incisors. Hooks were placed on the chains with elastics extending to a Delaire-type facial mask. Treatment is still underway

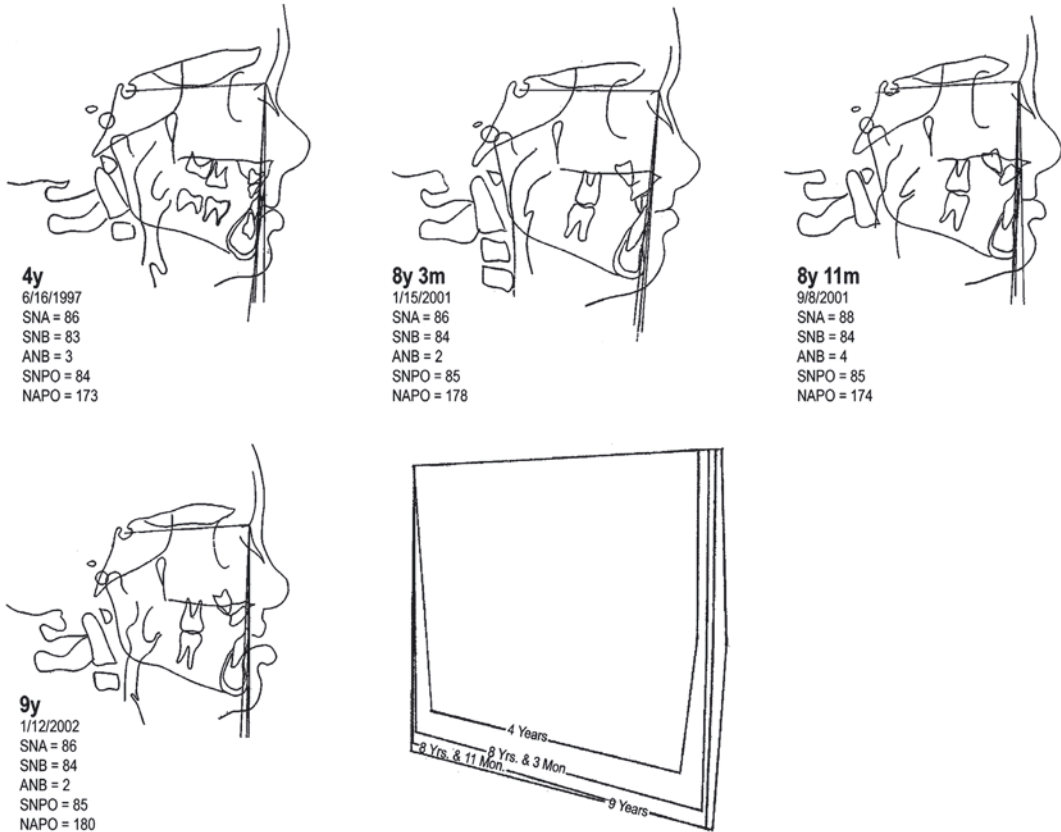
CASE # BT-80

Fig. 20.17. Lateral cephaloradiographs show the unerupted severely ventroflexed premaxilla in crossbite. The superimposed cephalometric tracing shows the continued midfacial growth keeping up with the other facial areas, resulting in a flat facial profile. One can conclude that growth at the premaxillary

vomerine suture is still active. However, the bony bridge between the lateral palatal segments and the premaxilla prevents the premaxilla's proper alignment and eruption into position



Fig. 20.18. Case BK 36. Bilateral cleft lip and palate. The protruding premaxilla was retruded and aligned within the arch using the POPLA procedure. At 7 years and 4 months of age an

anterior dental crossbite resulted. A protruding lower lip is due to the permanent mandible and flaring incisor teeth



Fig. 20.19.



Fig. 20.19. (continued) Case BK 36. Bilateral cleft lip and palate. The corrective treatment plan was designed to advance the crowded anterior maxillary incisors with facial protraction mechanics and position the cuspids into the missing lateral incisors spaces. Extraction of the mandibular first bicuspsids and retracting the incisors will help to reduce the crossbite without the need to advance the upper incisors. This case is still under treatment

CASE # BK-36

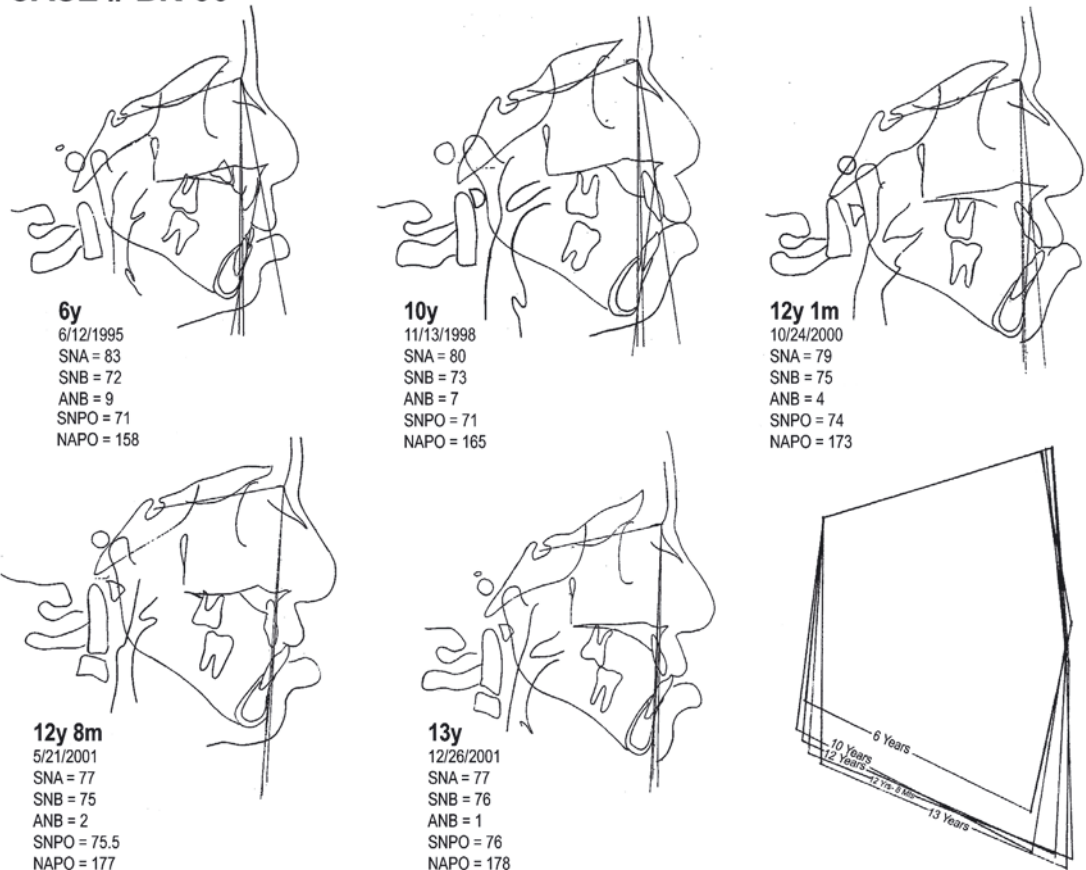


Fig. 20.20. Case BK 36. Bilateral cleft lip and palate. The premaxilla is POPLA retruded. The maxillary anterior teeth were gradually advanced but were still in crossbite. Superimposed cephaloradiographs show that forward mandibular growth enhanced the anterior crossbite; this necessitated the extraction

of the lower first bicuspsids and the retraction of the incisors. A proper overjet and overbite will be achieved when the mandibular incisors are retracted. A slight maxillary advancement of the incisors may still be necessary

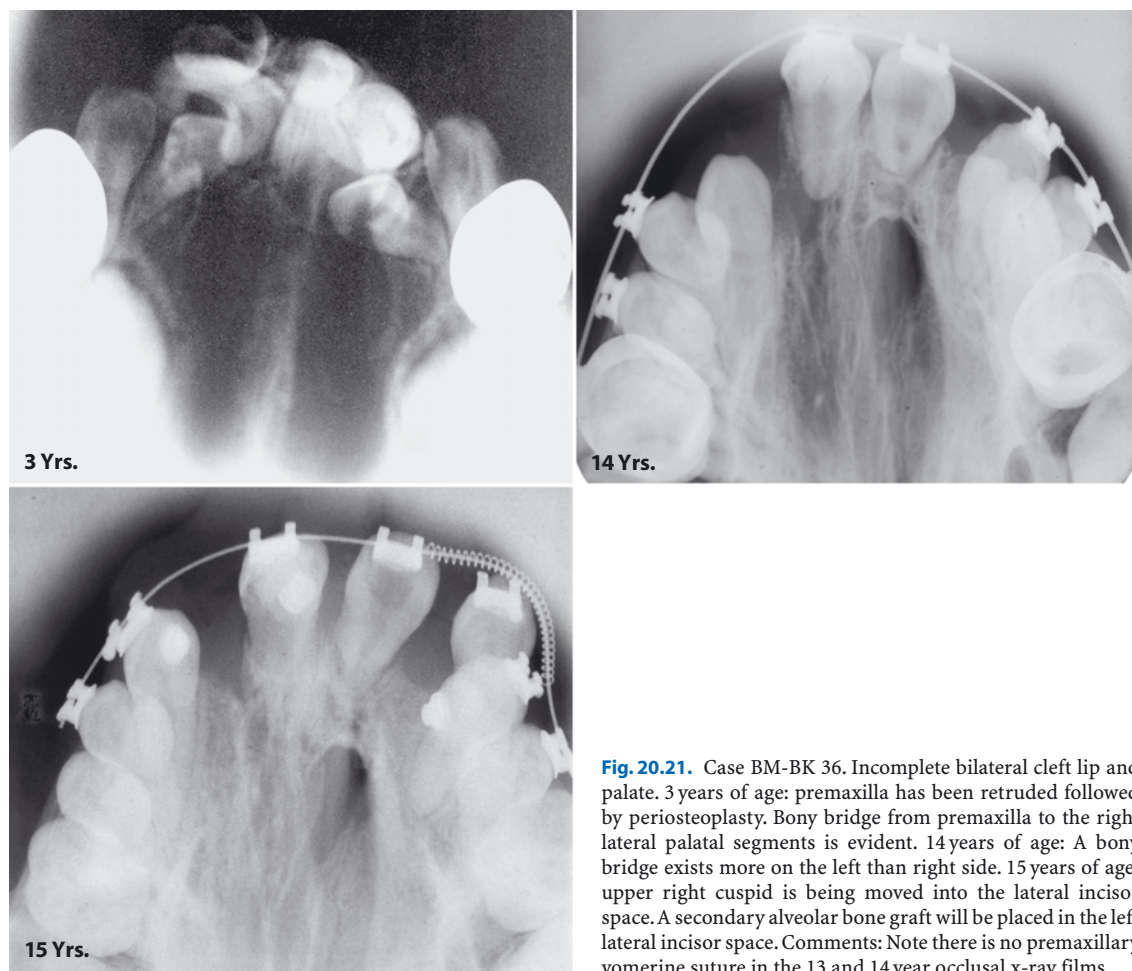


Fig. 20.21. Case BM-BK 36. Incomplete bilateral cleft lip and palate. 3 years of age: premaxilla has been retruded followed by periosteoplasty. Bony bridge from premaxilla to the right lateral palatal segments is evident. 14 years of age: A bony bridge exists more on the left than right side. 15 years of age: upper right cuspid is being moved into the lateral incisor space. A secondary alveolar bone graft will be placed in the left lateral incisor space. Comments: Note there is no premaxillary vomerine suture in the 13 and 14 year occlusal x-ray films

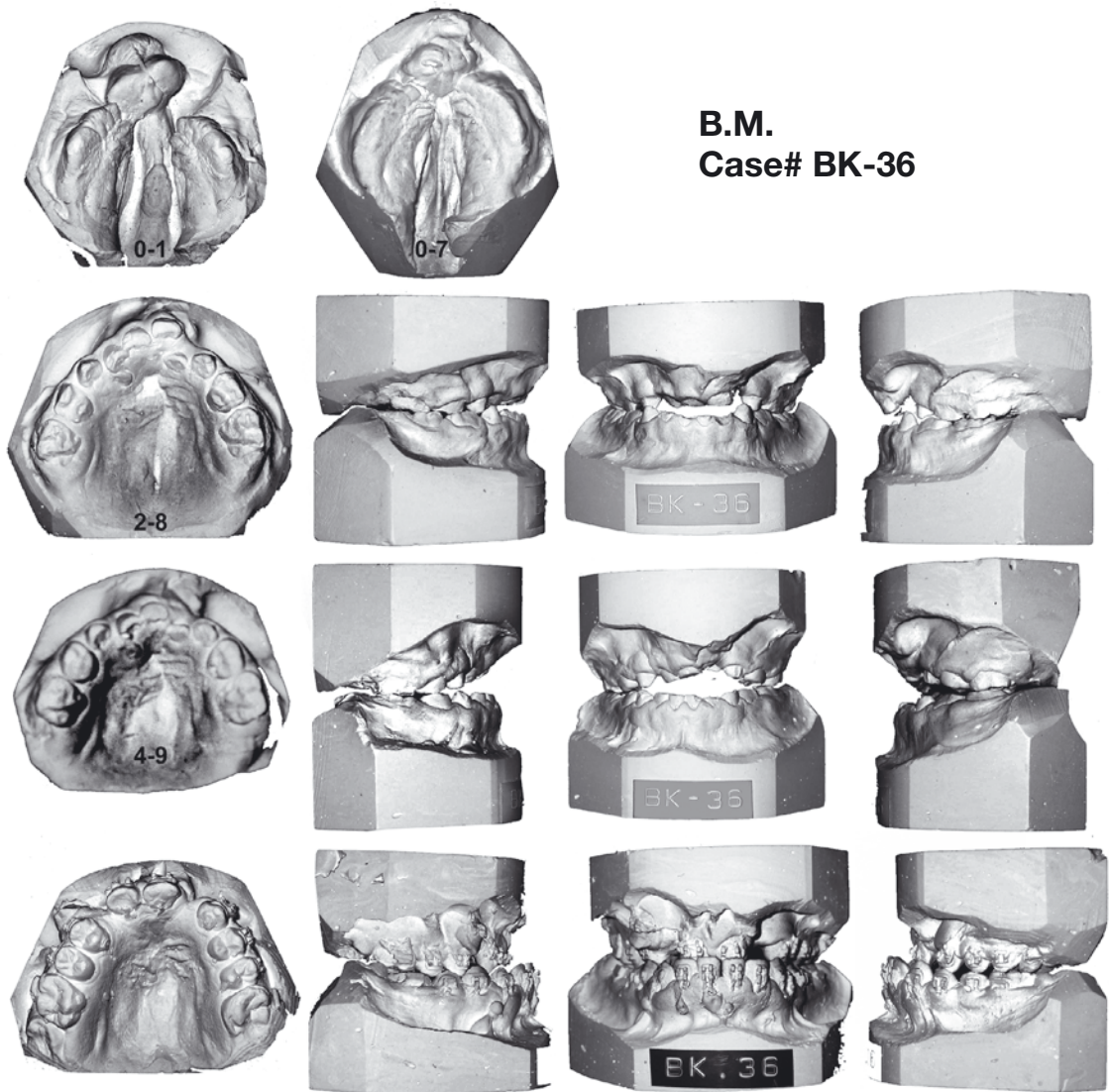


Fig. 20.22. Case BM-BK 36. Incomplete bilateral cleft lip and palate. 0-7: Untreated palatal arch. 2-8: The protruding premaxilla was treated with POPLA. 4-9: Class III malocclusion with an anterior open bite. 14-6: Missing right and left lateral incisors with closure of their spaces. Anterior-posterior arch length is shortened. Even after use of a protraction facial mask, extensive

orthodontics was still necessary to attempt to correct the amateur crossbite. Comments: Facial profile evaluation shows a protrusive lower lip with a severe mandibular incisor inclination. The lower first bicusps were extracted in order to retract the lower anterior teeth. The upper incisors will only be slightly advanced



Fig. 20.23 a–p. Case CS (AY-45) CUCLP. Latham-Millard PSOT. This case also demonstrates the loss of the lateral incisor space with slight midfacial retrusion at 9 years. **a** Newborn 3 months. **b** Intraoral view at 3 months. **c** 4 months after palatal manipulation; periosteoplasty, and lip adhesion. **d** Looking at

the periosteoplasty, palate cleft is open. **e** 9 months, after rotation advancement lip surgery. **f** 1 year, 3 months. Cleft palate closed via modified van Langenbeck procedure. **g** 2 years, 1 month. **h** 3 years, 8 months. **i** 5 years, 8 months



Fig. 20.23 a–p. (continued) **j** intraoral photograph with teeth in an anterior tip-to-tip relationship. **k, l** 7 years 8 months; frontal and lateral photographs. **m, n** 9 years, 10 months. Comment: The facial aesthetics are most pleasing even with the teeth in a tip-to-tip relationship. It still remains to be seen whether the profile will become more recessive after the pubertal growth spurt. **o** Final facial photographs at 18 years.

p Final occlusion at 18 years old showing good overbite and overjet. The upper left lateral incisor was replaced with PONTIC attached to a bridge. Comments: The results turned out well due to a vertical facial growth pattern

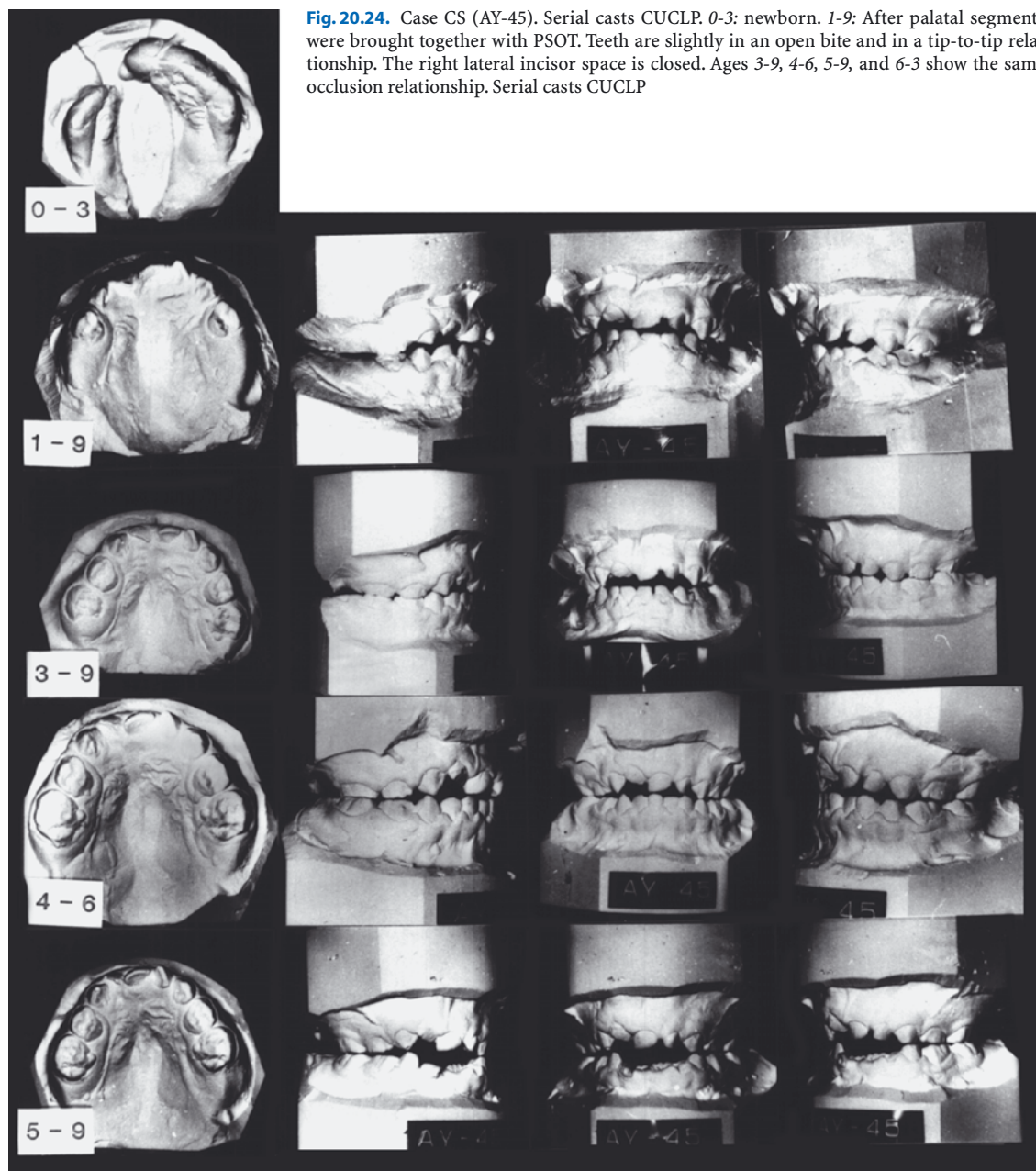


Fig. 20.24. Case CS (AY-45). Serial casts CUCLP. 0-3: newborn. 1-9: After palatal segments were brought together with PSOT. Teeth are slightly in an open bite and in a tip-to-tip relationship. The right lateral incisor space is closed. Ages 3-9, 4-6, 5-9, and 6-3 show the same occlusion relationship. Serial casts CUCLP

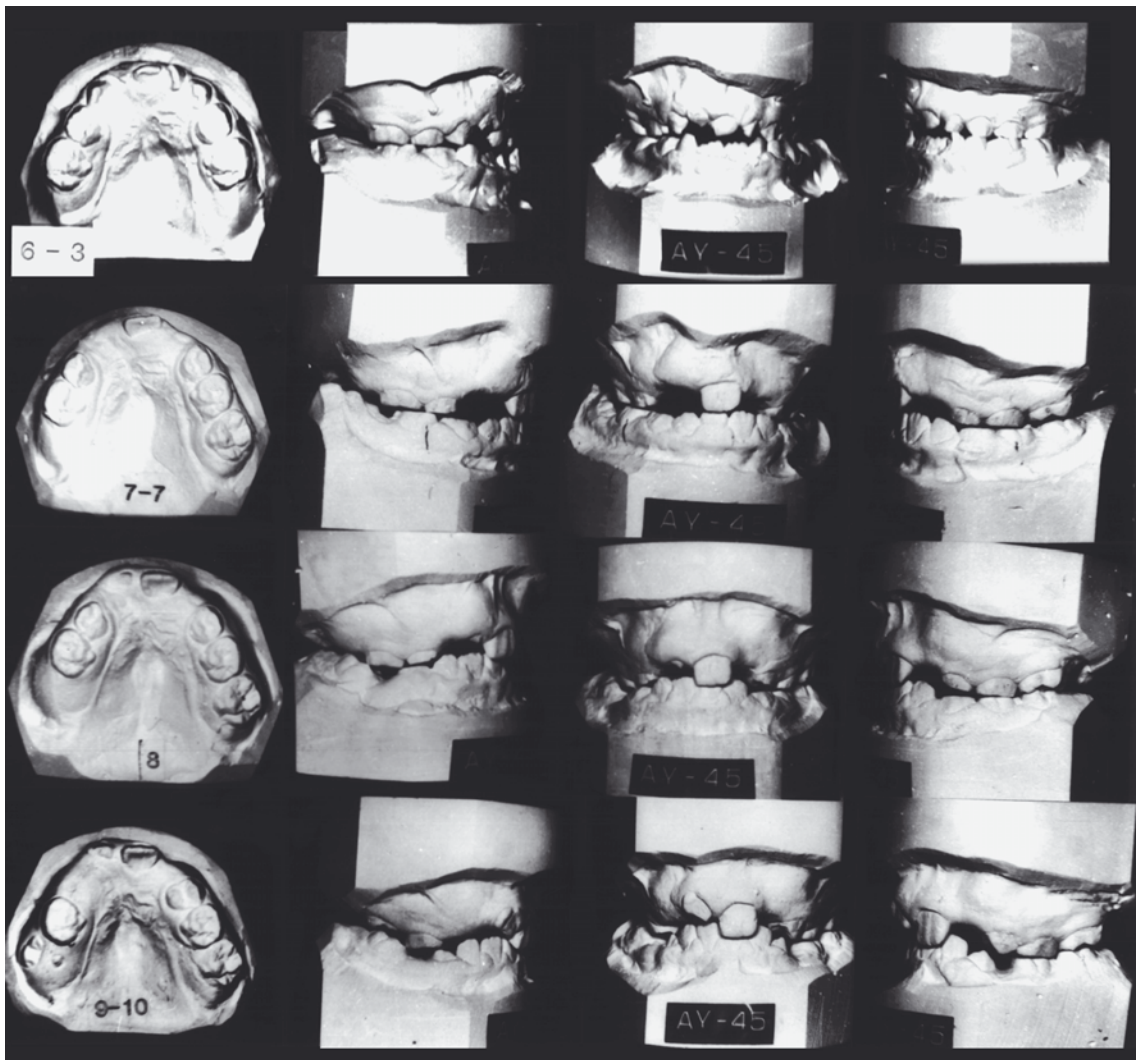


Fig. 20.24. (continued) At 7-7, 8, and 9-10: the permanent central incisors are erupting in good overjet-overbite relationship; however, the closure of the right lateral incisor space is due to

the maxillary incisors being displaced to the left. Serial casts CUCLP



Fig. 20.24. (continued) 10-9 to 13-0 Slight anterior crossbite. A protraction facial mask is to be used to advance the right central incisor. The left incisors are rotated and impacted. The lateral incisor is missing. 13 years: The left central incisor and

upper first bicusps were extracted. 16 years: A maxillary retainer with a false tooth was used. 18 years: A fixed bridge with replacement tooth was placed

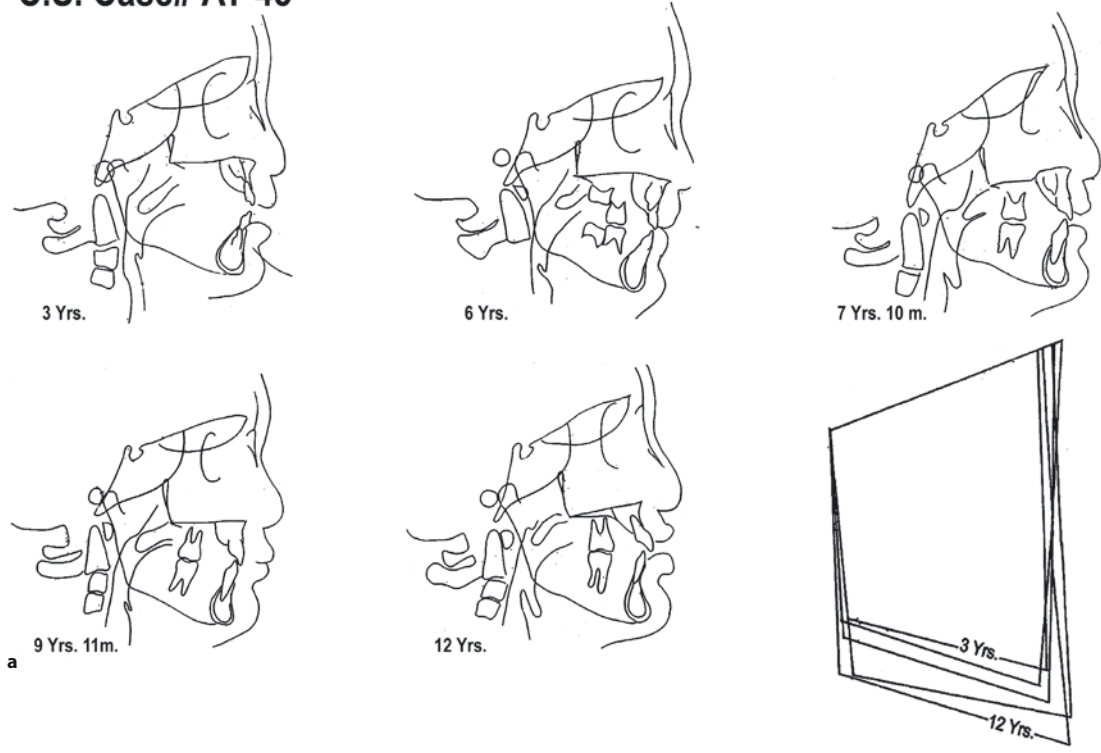
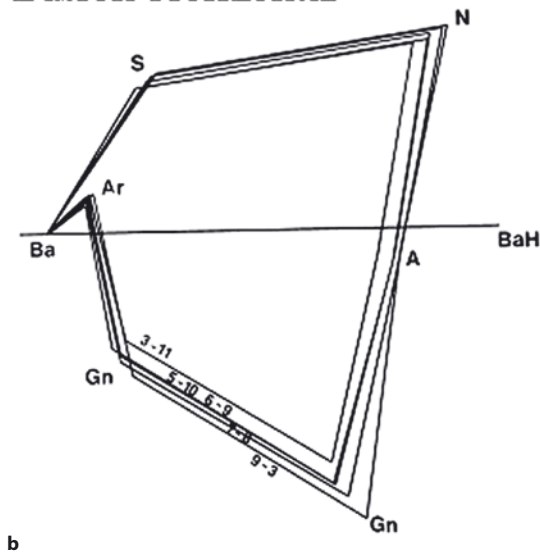
C.S. Case# AY-45**Basion-Horizontal**

Fig. 20.25 a,b. Case CS (AY-45). **a** Cephalometric tracings. Lateral cephalometric tracings show the midface is becoming more recessive relative to the anterior cranial base and mandible. **b** Superimposed polygons using Basion-Horizontal procedure. The above finding is verified in this series of superimposed polygons. Although slight forward growth of the midface is evident between 3-11 and 7-8, no further growth is seen between 7-8 and 9-3 years of age

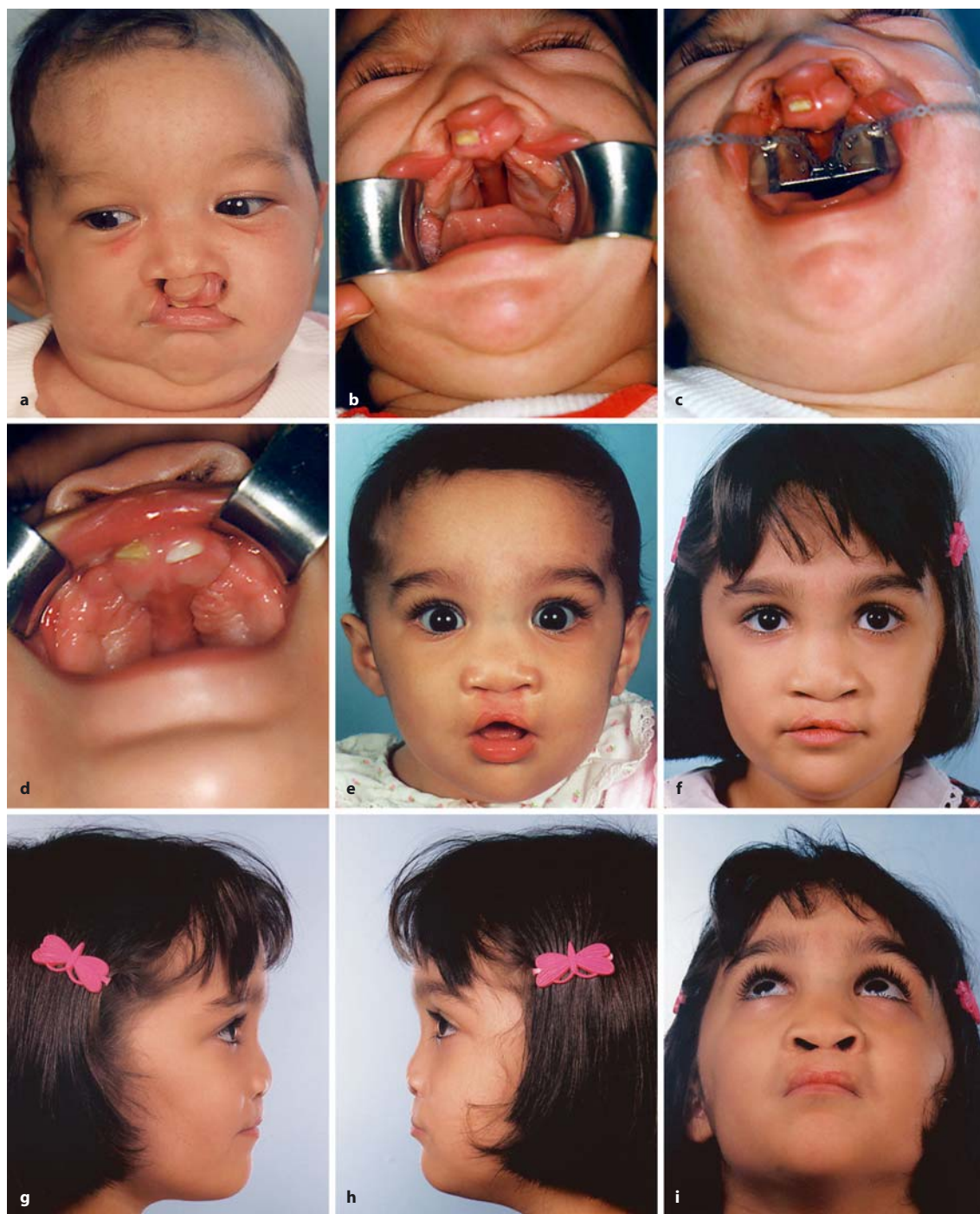


Fig. 20.26 a–w. Case AS (AY-46). CBCLP with Millard-Latham PSOT. Retarded midfacial growth in the mixed dentition. **a, b** Newborn. **c** Latham's pinned appliance in place. **d** At 6 months. Premaxilla is aligned within the arch. **e** At 6 months.

f–h 6 years and 6 months. Frontal and lateral face. **i** Small columella with depressed nasal tip. CBCLP with Millard-Latham PSOT



Fig. 20.26 a-w. (continued) **j** Good arch alignment. Small palatal fistula. **k** Good occlusion. **l-n** 9 years. Raised nasal tip. **o-r** Facial and dental occlusion. The left deciduous lateral inci-

sor is in crossbite. Retarded midfacial growth in the mixed dentition. CBCLP with Millard-Latham PSOT



Fig. 20.26 a–w. (continued) **s–u** Final facial photographs. **v, w** Final Occlusion

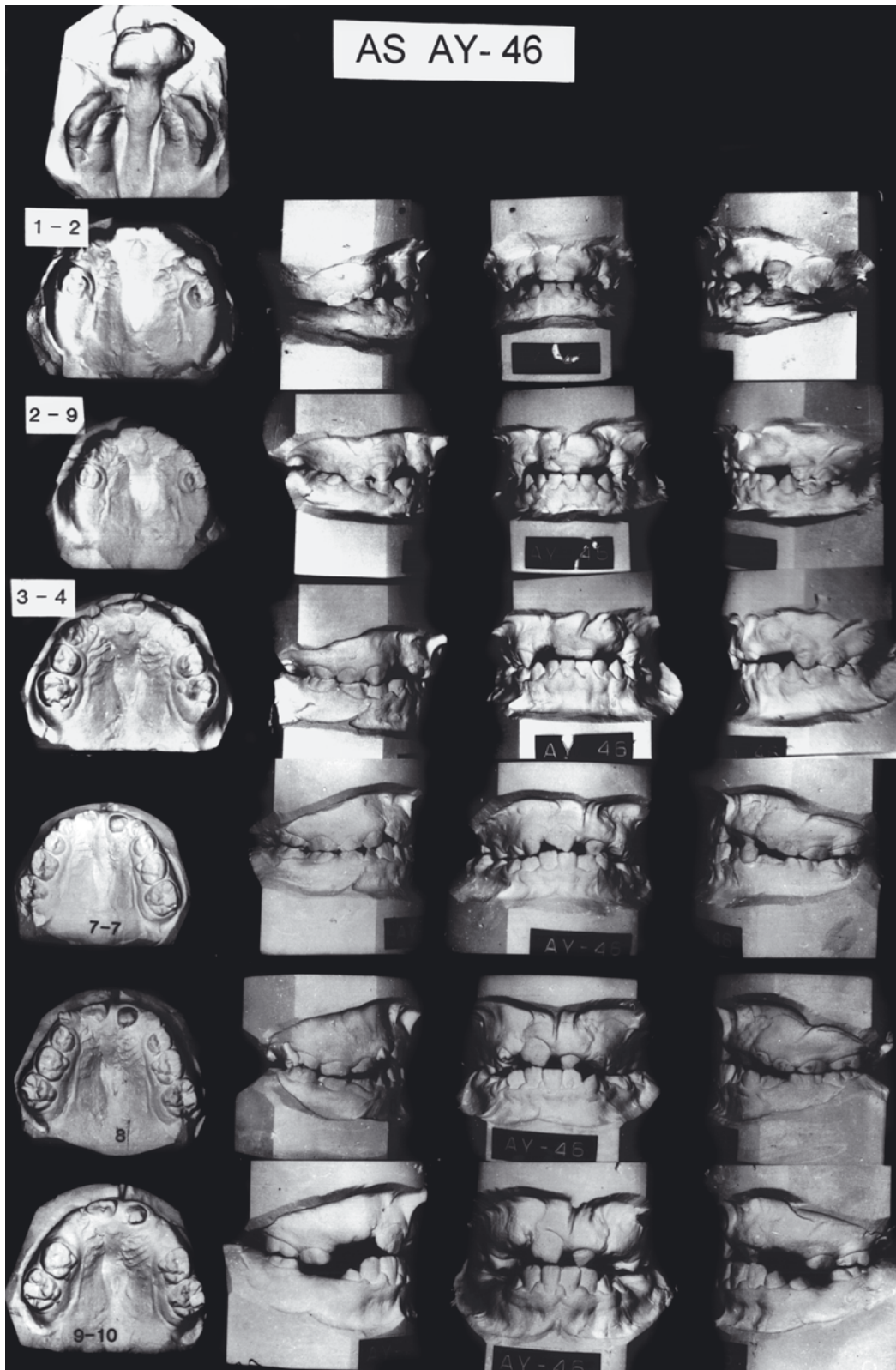


Fig. 20.27. Case AS (AY-46). CBCLP with Millard-Latham PSOT



Fig. 20.27. (continued) 10-9: Upper left lateral incisor is missing. The left maxillary central incisor is impacted. 18-0: Final occlusion. Treatment plan: 1. Extract upper left lateral incisor and bring in blocked out impacted central incisor. 2. Bring

upper right and left cuspids into lateral incisor position. 3. Reshape upper right cuspid. 4. Two-unit bridge to replace upper left lateral incisor

20.4 Conservative Non-POPLA CUCLP and CBCLP Cases

In non-POPLA CBCLP cases the protruding premaxilla's overjet is gradually reduced so that it can be easily aligned within the alveolar arch by 7–8 years of age. In some patients this may spontaneously occur after only 2–3 months of simple palatal expansion. However, depending on the facial growth pattern, some maxillary dental overjet with a convex facial profile may still remain into the mixed dentition stage. In our study only two cases with a severely protruding premaxilla at birth developed a retrusive Class III malocclusion requiring surgical maxillary LeFort I advance-

ment. This need was mainly due to reduced maxillary growth coupled with a forward-growing mandible. The use of a protraction facial mask in both cases was unsuccessful in correcting the midfacial recessiveness.

With non-POPLA treatments, successful secondary alveolar bone grafts occurred in 70% of the cases. In most of these cases, the impacted lateral incisor erupted into its normal position within the alveolus or, if absent, the space was left open for a replacement tooth. If one or both lateral palatal segments are in a Class II relationship and one or both lateral incisors are absent, the cuspid(s) can be substituted for the missing lateral incisor(s). In these patients the maxil-

A.S. Case# AY-46

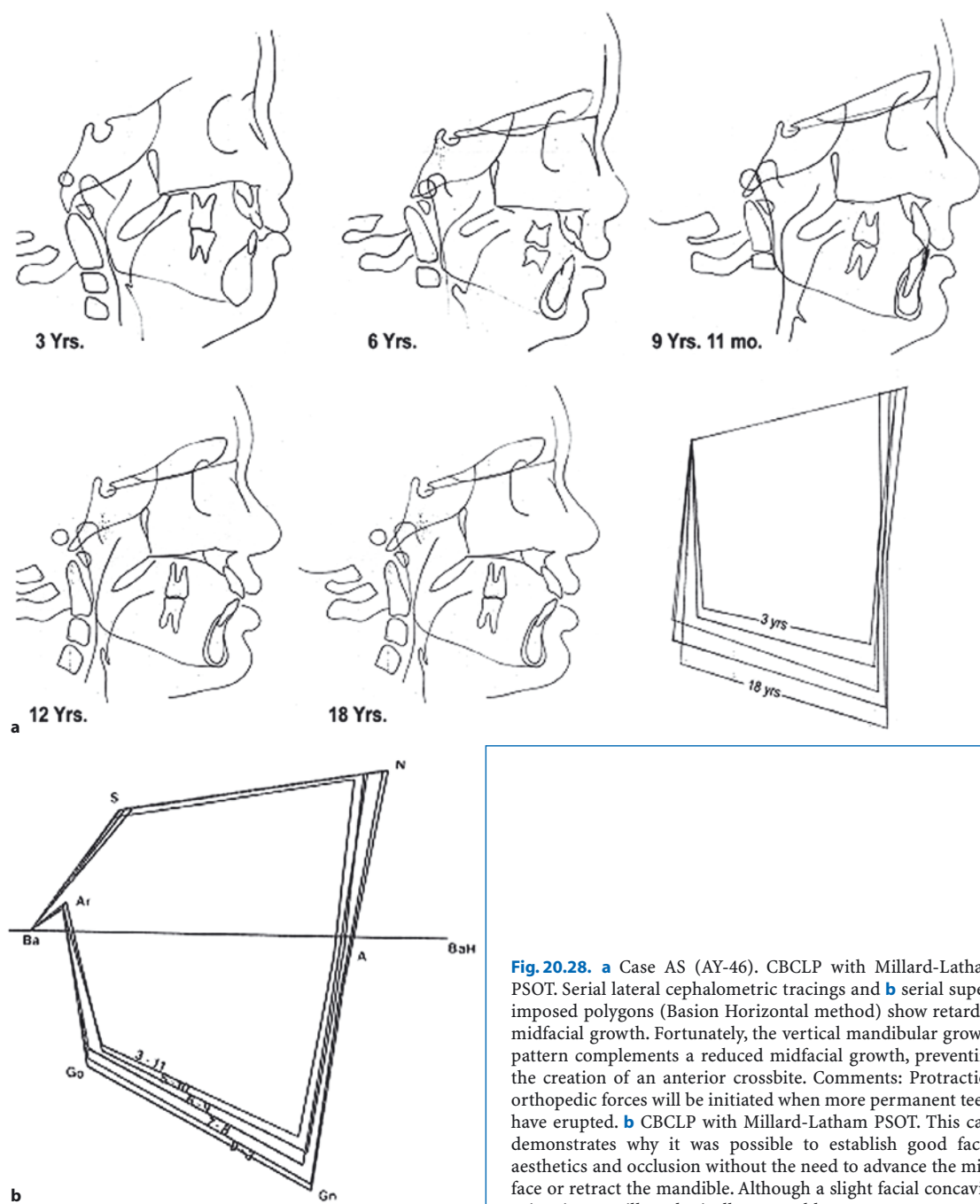


Fig. 20.28. **a** Case AS (AY-46). CBCLP with Millard-Latham PSOT. Serial lateral cephalometric tracings and **b** serial superimposed polygons (Basion Horizontal method) show retarded midfacial growth. Fortunately, the vertical mandibular growth pattern complements a reduced midfacial growth, preventing the creation of an anterior crossbite. Comments: Protraction orthopedic forces will be initiated when more permanent teeth have erupted. **b** CBCLP with Millard-Latham PSOT. This case demonstrates why it was possible to establish good facial aesthetics and occlusion without the need to advance the mid-face or retract the mandible. Although a slight facial concavity exists, it was still aesthetically acceptable

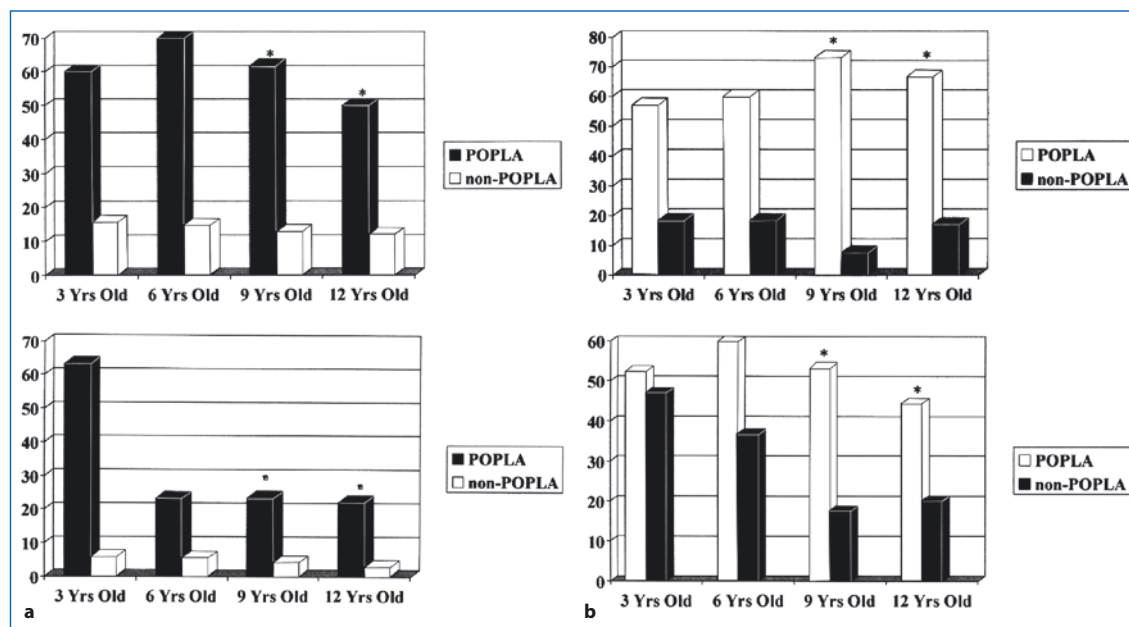


Fig. 20.29. **a** The charts report the percentage of children with unilateral cleft lip and palate receiving presurgical or nonpresurgical orthopedics, by crossbite. At every age level, a greater percentage of youngsters treated with presurgical orthopedics developed crossbites than did those treated with nonpresurgical orthopedics (presurgical orthopedics, nonpresurgical orthopedics). This difference existed for both the anterior crossbite (*above*) and the buccal crossbite (*below*). Asterisk: some of the worst crossbite cases in the presurgical orthopedics group were treated by Berkowitz starting at about 9 years of age, thereby reducing the frequency of crossbite in this group beginning at this age level. **b** The charts report the

percent of children with bilateral cleft lip and palate receiving presurgical orthopedics or nonpresurgical orthopedics, by crossbite. At every age level, a greater percentage of youngsters treated with presurgical orthopedics developed crossbites than did those treated with nonpresurgical orthopedics (presurgical orthopedics; nonpresurgical orthopedics). This difference existed for both the anterior crossbite (*above*) and the buccal crossbite (*below*). Asterisk: some of the worst crossbite cases in the presurgical orthopedics group were treated by Berkowitz starting at about 9 years of age, thereby reducing the frequency of crossbite in this group beginning at this age level

lary and mandibular anterior arch forms are congruent at the completion of orthodontic treatment. Anterior arch congruency cannot exist in POPLA cases without extensive orthodontics/surgery.

Flattening of the facial profile, thereby contributing to improved facial aesthetics, occurs gradually as the upper and lower portions of the face grow forward while the midface's forward growth is restrained by the forces created by the united lip. Most significantly, after 2 years of age 3D measurements show that the premaxilla is in the same ideal position within the maxillary arch, its forward growth having been retarded by the intact lip musculature forces.

Poor facial growth occurs either when the mandible's growth is vertically directed and/or the midface fails to grow in concert with the mandible. Any of these variables can occur in either non-POPLA or POPLA treated cases which are not under the surgeon's control, and are unpredictable at birth.

The growth analysis of these CBCLP cases demonstrates that increases in A-P maxillary arch length,

between the incisal papilla and the first permanent molars, is dependent on the functional integrity of the PVS. Once PVS function is diminished by excessive forces growth recovery will not return.

Establishing lip muscle continuity soon after birth causes slow molding of both the overexpanded lateral palatal and premaxillary segments leading to the narrowing of the anterior and posterior palatal cleft spaces. In non-POPLA CBCLP the posterior-directed pressure of the elastic strip appears to be within physiological limits and in most cases reduced pressure permits additional PVS growth. This is seen in the graphic analysis of both superimposed serial lateral cephaloradiographs and from electronically produced 3D tracings of palatal dental casts. (Figs. 20.30, 20.31.)

In non-POPLA CBCLP cases, the A-P palatal dimension is greater between birth and 6 years, while with POPLA CBCLP treatment, once the premaxilla is set back there is no further change in the antero-posterior first molar to incisor dimension due to pres-

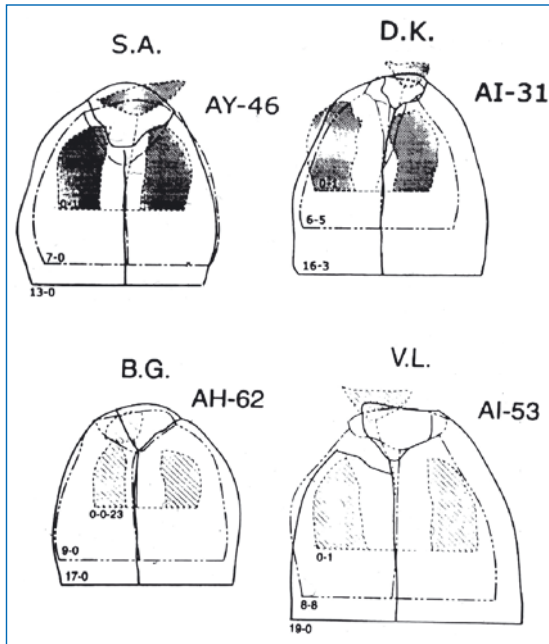


Fig. 20.30. Superimposed palatal cast tracings of the hard palate of four patients were acquired using a 3D electro-mechanical digitizer. In each tracing, the alveolar ridge is the lateral border. The tracings are superimposed horizontally at the rugae and registered vertically

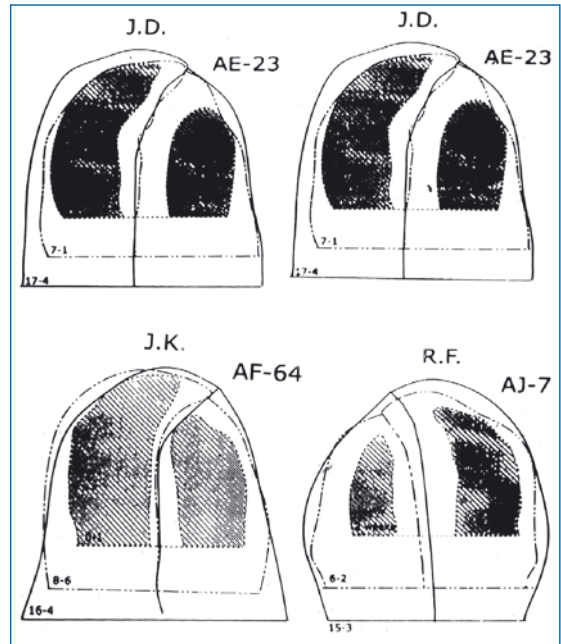


Fig. 20.31. Superimposed palatal cast tracings of the hard palate of four patients were acquired using a 3D electro-mechanical digitizer. In each tracing, the alveolar ridge is the lateral border. The tracings are superimposed horizontally at the rugae and registered vertically

Table 20.2. Obtained significant chi-square values ($p < 0.05$) in tests of differences in number of patients having crossbites between presurgical and nonpresurgical orthopedics treatments

	Approximate age of participant				
	3 years	6 years	9 years	12 years	Total sample
Unilateral cleft					
Anterior crossbite	16.8	30.0	10.2	18	77.5
Buccal crossbite	30.9	6.5	6.5	4.8	42.8
Bilateral cleft					
Anterior crossbite	10.3	11.5	24.5	8.5	35.5
Buccal crossbite	49	49	6.5	35	173

sure-caused hemorrhage at the premaxillary vomerine suture leading to fibrosis and then synostosis. Most of the alveolar cleft spaces are obliterated with new bone uniting the repositioned palatal segments.

In the non-POPLA CUCLP cases studied, only 16% (8 of 51) are in anterior crossbite at 3 years of age, a condition that is easily correctable by advancing the premaxillary portion of the noncleft larger segment. 60% of the POPLA cases are anterior crossbites at 3 years. (Fig. 20.29a)

Buccal crossbites in both non-POPLA and POPLA treatment series were easily corrected since the type of palatal surgery performed between 18 and 36 months did not inhibit palatal expansion. Only 3 of 51 cases (6%) had a complete buccal crossbite at 3 years of age (Table 20.1, Fig. 20.29a). The relative increase in percentage of buccal crossbites in POPLA cases at 3 years is significant, (60% at 3 years) because it necessitated more crossbite corrective procedures at 5–6 years of age (Table 20.2, Fig. 20.29a).

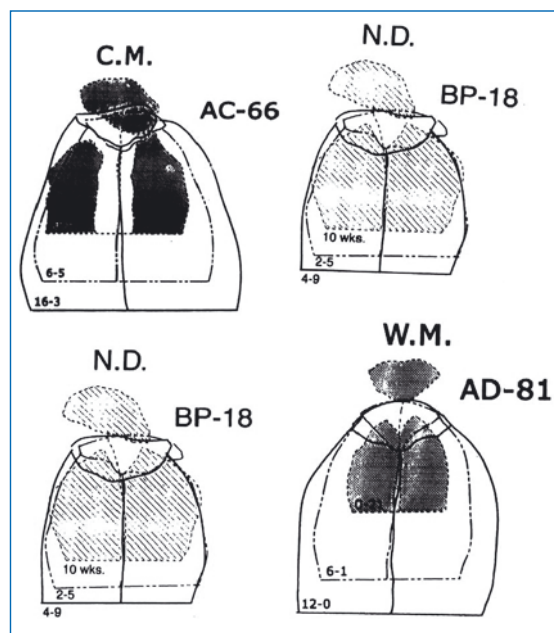


Fig. 20.32. Superimposed palatal cast tracings of the hard palate of four patients were acquired using a 3D electro-mechanical digitizer. In each tracing, the alveolar ridge is the lateral border. The tracings are superimposed horizontally at the rugae and registered vertically

20.5 Variations in Palatal Osteogenic Deficiency and Its Influences on Surgical Treatment

The location of palatal bone deficiency is highly variable and can exist to various degrees. In CBCLP the extent of osteogenic deficiency in the anterior part of the palate can vary in size and shape. In a few of these instances the palatal cleft space is usually present immediately posterior to the premaxilla, even when the incisors are in good or poor overbite/overjet relationship. In conservatively treated cases, if this large cleft space is present with the incisors in good overbite/overjet, further surgical treatment needs to be evaluated in the mixed dentition or at early adolescence. When there is insufficient mucoperiosteal tissue to surgically close this palatal cleft space, successful secondary alveolar bone grafting will not be possible. In these instances the only solution is for the posterior palatal segments to be advanced to reduce the cleft space to usable dimensions and immediately followed by secondary alveolar bone grafts [29]. In our study this surgery was necessary in only two of 20 non-POPLA cases; both cases were successfully treated by Wolfe using the Posnick procedure.

Should this same form of skeletal palatal deficiency exist in POPLA-treated cases, the premaxilla would be severely retruded, so much so that one or both lateral incisor cleft spaces would be closed and ultimately bridged by bone, thus preventing the orthodontic-orthopedic correction of the anterior crossbite. The lack of subsequent anterior maxillary and mandibular arch congruency would prevent the achievement of a good dental overbite/overjet relationship, even after midfacial surgical advancement.

20.6 Correction of Midfacial Deficiencies in Conservatively Treated Non-POPLA Cases

A slight anterior crossbite can be easily corrected orthodontically by using the maxillary protraction mechanics of a facial mask, with or without the extraction of one mandibular central incisor. In the non-POPLA CBCLP cases it was necessary to surgically advance a retrusive midface in only two patients. A well-developed, relatively protrusive mandible existed in only one of these children. Fortunately, no long-term speech changes resulted after maxillary advancement in these two instances. Lateral cephalometrics showed good velar length and elevation within a pharyngeal space of average depth. Intraoral examination showed good lateral pharyngeal wall movements as well.

20.7 Similar Presurgical Orthopedics As It Was Utilized in the Past – It Failed Then As It Does Now

During the 1920s and 1930s the cleft surgeon's treatment philosophy exemplified by Brophy [30] was to repair the cleft defect by establishing "normal" anatomical palatal form soon after birth, through the use of external and internal palatal compression techniques. The first priority then was to improve facial aesthetics, followed in turn by good dental function and speech. Unfortunately, the Brophy procedure led to extensive midfacial deformity and was eventually discontinued.

Due to the benefit of long-term facial and palatal growth records, many if not most surgeons and speech-language pathologists have recognized the extent to which time, or more precisely growth, serves as their ally or enemy. However, some surgeons are still endeavoring to devise a procedure that can be used during the first 2 years for all CBCLP and CUCLP cases, hopeful that good facial growth will follow.

In recent years, serial documentation of the natural evolution of postnatal facial and palatal development

of children with complete bilateral and unilateral cleft lip and palate has yielded important objective data – data that help explain the dynamics of facial skeleton and palate growth under the influence of various surgical procedures. This knowledge has greatly improved the ability of surgeons and orthodontists to develop physiologically based concepts that lead to successful long-term treatment outcomes.

This study supports Gillie's (personal communication with Millard) belief that time is both the surgeon's ally and most trenchant critic. It further supports the thesis that: No single surgical procedure performed at birth is suitable for all cleft types and faces since there is great variation in palatal osteogenesis and in facial growth patterns. Staged treatment based on the individual patient's facial assets and deficits must be the controlling factor in designing therapy.

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21 Nasoalveolar Molding for Infants Born with Clefts of the Lip, Alveolus and Palate

BARRY H. GRAYSON, DEIRDRE MAULL

21.1 Introduction

Cleft lip and palate can present with considerable variation in severity and form. Generally, the wider, more extensive clefts are associated with more significant nasolabial deformity. These clefts, deficient in both hard and soft tissue elements, present a significant surgical challenge to the achievement of a functional and cosmetic outcome. Even a mild incomplete unilateral cleft lip in the absence of a cleft palate can be associated with a nasal deformity. Most surgeons would agree that their chance of achieving a finer surgical scar, good nasal tip projection, and a more symmetrical and precisely defined nasolabial complex would be better in an infant that presents with a minor cleft deformity. It is traditional surgical experience that a finer scar forms when a surgical incision heals under less rather than more tension. The principle objective of presurgical nasoalveolar molding is to reduce severity of the initial cleft deformity. This enables the surgeon and ultimately the patient to enjoy the benefits associated with repair of cleft deformity that is of minimal severity.

The unilateral cleft deformity is characterized by a wide nostril base and separated lip segments on the cleft side. The affected lower lateral nasal cartilage is displaced laterally and inferiorly, which results in a depressed dome, the appearance of an increased alar rim, an oblique columella, and an overhanging nostril apex. If there is a cleft of the palate, the nasal septum will deviate to the noncleft side with an associated shift of the nasal base [7].

The bilateral cleft lip deformity typically presents with a procumbent and/or rotated premaxilla. The alar base width is significantly increased and the lip segments are widely separated [8]. The flattened nasal tip appears tethered directly to the prolabium by a severely deficient or absent columella. The lower lateral alar cartilages are flared and concave where they should be convex. Certainly, the greatest challenge for

esthetic reconstruction is the absent or deficient columella.

21.2 History

There have been numerous techniques documented over the centuries to improve the position of the cleft alveolar segments. In 1686, Hoffman described the use of a head cap with arms extended to the face to retract the premaxilla and narrow the cleft. There have been many improvements to this method of using the head as extraoral anchorage [5], and it is still used today to retract the premaxilla [1]. The concept of an intraoral device to reposition the cleft alveolar segments is attributed to McNeil [3]. Through a series of acrylic plates, the segments were actively molded into the desired position. In 1975, Georgiade [4] and Latham introduced a pin-retained appliance to simultaneously retract the premaxilla and expand the posterior segments over a period of days. In response to controversy associated with active retraction of the premaxilla, Hotz described the use of a passive orthopedic plate to slowly align the cleft segments [2]. The premaxilla is not retracted, and by age 10, Hotz feels that the face has grown forward into appropriate balance with the premaxilla. All of these appliances were designed to correct the alveolar cleft only, despite the fact that the cleft nasal deformity remains the greatest esthetic challenge.

In 1993, Grayson et al. [9] described a technique to correct the alveolus, lip, and nose in infants born with cleft lip and palate. The original research for molding cartilage was performed by Matsuo [18–21]. He recognized that the cartilage in the newborn is soft and lacks elasticity. He believed that the high maternal level of estrogen at the time of birth correlates with an increase in hyaluronic acid, which inhibits the linking of the cartilage intercellular matrix. This process may be necessary to relax ligaments, cartilage, and connec-

tive tissue, enabling the fetus to pass through the birth canal. The level of estrogen begins to decline immediately after birth. Matsuo used a stent in the form of a pair of silicone tubes to shape the nostrils, with some limitations. These include the need for an intact nasal floor, (Simon Arts band or lip adhesion) and the inability to direct the force since his stent expands circumferentially. Grayson adapted his nasal stent to extend from the anterior flange of an intraoral molding plate. This new technique was called nasoalveolar molding (NAM) [15–17]. The greatest advantage of this technique is that it enables the practitioner to apply force to precisely shape the nasal cartilage and in the case of bilateral cleft lip and palate, lengthen the columella. In addition, since the stent is extended from a molding plate, an intact nasal floor is not required.

21.2.1 Objectives

The principle objective of presurgical nasoalveolar molding is to reduce severity of the initial cleft deformity. This enables the surgeon to enjoy the benefits associated with repair of an infant that has a minimal

cleft deformity. These goals include lip segments that are almost in contact at rest, symmetrical lower lateral nasal cartilages, uprighting of the columella and adequate nasal mucosal lining that permits postsurgical retention of the projected nasal tip.

Additional objectives of nasoalveolar molding include reduction in the width of the alveolar cleft segments until passive contact of the gingival tissues is achieved. As reduction of the alveolar gap width is accomplished, the base of the nose and lip segments achieve improved alignment.

Tapes that actively bring the lip segments together are used in conjunction with the molding plate and nasal stent. Taping the lips together helps to upright the inclined columella along the midsagittal plane. As the lower midface skeletal elements (alveolar ridges and lower maxilla) improve relationship, the overlying soft tissues follow (Fig. 21.1 a–d).

As reduction of alveolar gap width is accomplished, the base of the nose and lip segments achieve improved alignment. As the alveolar ridges and lower maxilla improve their relationship, the overlying soft tissues improve. The alar rim, which was initially stretched over a wide alveolar cleft deformity, shows some laxity, which enables it to be elevated into a sym-



Fig. 21.1. **a** Patient before the implementation of nasoalveolar molding. Note the wide nostril base and widely separated lip segments. The affected lower lateral nasal cartilage is displaced laterally and inferiorly, which results in a depressed dome, the appearance of an increased alar rim, an oblique columella, and an overhanging nostril apex. **b** Patient following nasoalveolar molding, just before primary surgical repair. Note the marked reduction in severity of cleft deformity, improving the prognosis of surgical outcome. The lip segments are in closer approximation at rest, uprighting of the columella, convexity of the lower lateral alar cartilage and projection of the nasal tip. **c** Frontal view and **d**, base view of patient at age 2 years, 5 months showing a minimally detectable lip scar good nasolabial esthetics



Fig. 21.2. Unilateral nasal stent in position showing lip taping and correction of nasal form. As reduction of alveolar gap width is accomplished, the base of the nose and lip segments achieve improved alignment. As the alveolar ridges and lower maxilla improve their relationship, the overlying soft tissues improve. The alar rim, which was initially stretched over a wide alveolar cleft deformity, now shows some laxity, which enables it to be elevated into a symmetrical and convex form. The nasal tip on the cleft side is overcorrected in its projection. This is achieved through the use of a nasal stent, intraoral molding plate, and surgical tape



Fig. 21.3. Infant feeding with bilateral nasoalveolar molding plate in place. Parents are instructed to keep the plate in full time, and to take it out for cleaning as needed, at least once a day. Initially, it may take longer to feed the infant with the plate in place, but the child quickly adjusts and parents report that they soon will not eat without it

metrical and convex form. The nasal tip on the cleft side is overcorrected in its forward projection. This is achieved through the use of a nasal stent, intraoral molding plate, and surgical tape (Fig. 21.2).

In the infant with bilateral clefts of the lip, alveolus, and palate, the objective of presurgical nasoalveolar molding includes the nonsurgical elongation of the columella, centering of the premaxilla along the mid-sagittal plane, and retraction of the premaxilla in a slow and gentle process to achieve continuity with the posterior alveolar cleft segments. Additional objectives include reduction in the width of the nasal tip, improved nasal tip projection, and decrease in the width of nasal alar base.

21.3 Procedure

A heavy-bodied impression material is used to take the initial impression as soon after birth as possible. We begin losing valuable time for the cartilage to remain plastic and moldable after the baby is born. In case of an airway emergency, the surgeon is always present to help with the impression. The infant is held upside down by the surgeon when the impression tray is inserted into the oral cavity. The tray is seated until impression material is observed just beginning to extrude past its posterior border. The infant is kept in the inverted position to keep the tongue forward and

to allow fluids to drain out of the oral cavity. Once the impression material is set, the tray is removed and the mouth is examined for residual impression material that may be left behind. A cast or model of the alveolar anatomy is made by filling the impression with a dense plaster material (dental stone). The molding plate is fabricated on the dental stone model. It is made of hard clear acrylic and lined with a thin coat of soft denture material. Care is taken to reduce the border of the plate in the area of the labial frenum attachments and other areas that may be likely to ulcerate. A 5-mm diameter hole is made in the center of the acrylic palatal vault to provide an airway in the event that the posterior border of the plate drops down onto the tongue. Parents are instructed to keep the plate in full time, and to take it out for cleaning as needed, at least once a day. Initially, it may take longer to feed the infant with the plate in place, but the child quickly adjusts and parents report that they soon will not eat without it (Fig. 21.3). The appliance is secured extraorally to the cheeks, bilaterally by surgical tapes, which have an orthodontic elastic band at one end. The elastics loop over a retention arm extending from the anterior flange of the plate (Fig. 21.4a,b). The retention arm is positioned approximately 40° down from the horizontal plane to achieve proper activation and to prevent unseating of the appliance from the palate. The tapes and elastics are changed once a day.

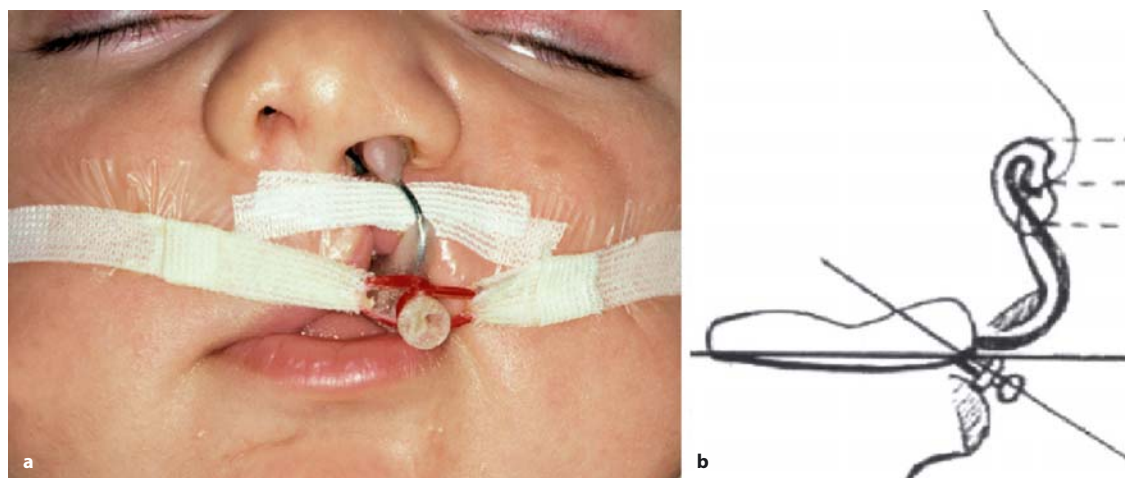


Fig. 21.4. **a** Unilateral nasal stent in position showing lip taping. The appliance is secured extraorally to the cheeks, bilaterally by surgical tapes, which have an orthodontic elastic band at one end. The elastics loop over a retention arm extending from

the anterior flange of the plate. **b** The retention arm is positioned approximately 40° down from the horizontal plane to achieve proper activation and to prevent unseating of the appliance from the palate

Weekly visits are required to modify the molding plate in order to guide the alveolar cleft segments into the desired position. Closure of the alveolar gap brings the lip segments together, reduces the nasal base width and introduces laxity of the alar rim. Care should be taken not to add the nasal stent before achieving laxity of the alar rim, as an increase of the nostril circumference may result.

Only one retention arm is attached to the appliance for treatment of the unilateral cleft patient. To determine its location on the labial border of the molding plate, the cleft lip segments are pulled together while centering the philtrum and columella. A pencil mark is placed on the labial flange of the molding plate at the junction of the cleft lip segments. The retention arm is attached at this point. The vertical position of the retention arm should be at the junction of upper and lower lips at rest. This will allow for approximation of the cleft lip segments and will not interfere with the resting position of the lower lip.

When the retention arms are engaged by the tape-elastic system, the elastics (inner diameter 1/4 in, wall thickness: heavy) should be stretched approximately two times the resting diameter for proper activation force (2 oz). The amount of force may vary depending on the clinical objective and the mucosal tolerance to ulceration. Retraction of the premaxilla will require greater elastic traction force than is required for closure of a unilateral alveolar gap.

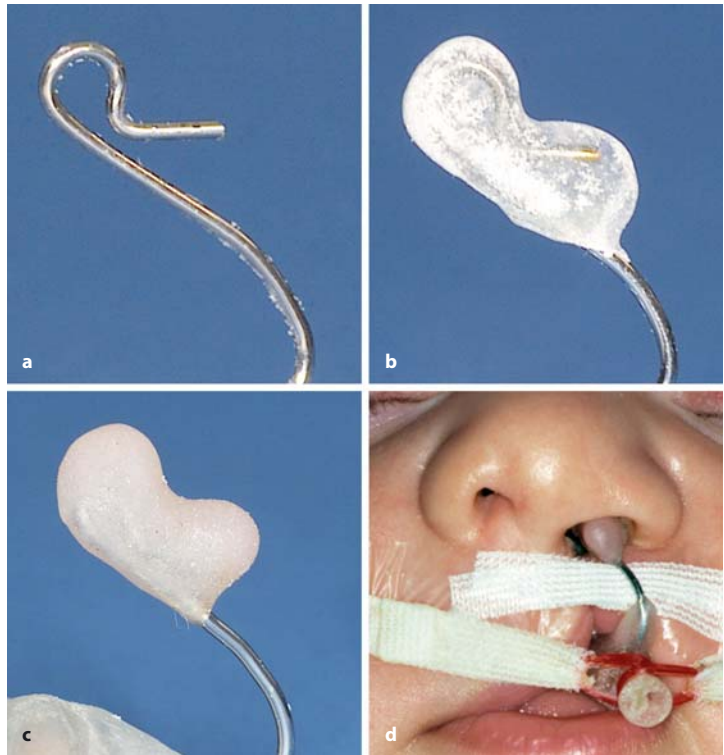
Parents are instructed to place tapes in order to approximate the cleft lip segments. The tape should be applied at the base of the nose (nasolabial angle) and



Fig. 21.5. The nasal stent is added to the intraoral molding plate when the cleft alveolar gap is reduced to 5 mm or less. The stent armature is made of .036 gauge round stainless steel wire. The intranasal portion is formed from hard acrylic that is covered with a thin layer of soft spongy acrylic denture liner

not low on the lip near the vermilion border. Taping too low can cause undesirable horizontal lengthening of the lip over time. The tape should be applied to the noncleft side first, then pulled over and adhered to the cleft side, making an effort to bring the philtrum and columella to the midline. Lip taping provides some of the benefits of a surgical lip adhesion without the associated surgical morbidity, hospital admission, cost, and scarring.

Fig. 21.6. **a** As the wire armature of the nasal stent extends into the nostril, it is curved back on itself to create a small loop for retention of the intranasal portion of the nasal stent. **b** Hard acrylic (methyl-methacrylate) is applied to the wire armature and shaped into a bi-lobed form that resembles a kidney. **c** The hard acrylic nasal stent is coated with a thin layer of soft denture liner for comfort. **d** The upper lobe of the nasal stent enters the nose and gently lifts the dome until a moderate amount of tissue blanching is evident. The lower lobe of the nasal stent lifts the nostril apex and defines the top of the columella



When the cleft alveolar gap is reduced to 5 mm or less, the nasal stent is added. The stent is made of .036 gauge round stainless steel wire and takes the shape of a “swan neck” (Fig. 21.5). A template for the nasal stent is made from a malleable rope of soft dental wax. The wax rope is attached to the labial flange of the molding plate, near to the base of the retention arm. It extends forward then curves backward (in the form of a swan neck) entering 3–4 mm past the nostril aperture. By copying the shape of this wax template one can then easily form the wire armature of the nasal stent. As the wire extends into the nostril, it is curved back on itself to create a small loop for retention of the intranasal portion of the nasal stent. The intranasal portion consists of a hard acrylic component which is shaped into a bi-lobed form resembling a kidney. A layer of soft denture liner is added to the hard acrylic for comfort. The upper lobe enters the nose and gently lifts the dome until a moderate amount of tissue blanching is evident.

The lower lobe of the nasal stent lifts the nostril apex and defines the top of the columella (Fig. 21.6a–e).

In the patient with bilateral clefts, there is a need for two retention arms and nasal stents. The fabrication steps are the same as described for the unilateral cleft. Each nasal stent originates from the molding plate at the base of a retention arm. After the nasal stents are added, attention is focused on nonsurgical lengthening of the columella. To achieve this objective a horizontal band of soft denture material is added to join the left and right lower lobes of the nasal stents, spanning the base of the columella. This band sits at the nasolabial junction and will define this angle as the nasal tip continues to be lifted and projected forward. Tape is adhered to the prolabium, underneath the horizontal lip tape, and stretches downward to engage the retention arms with elastics. This vertical pull is a counter-stretch to the upward force applied to the nasal tip by the nasal stent. Taping downward on the prolabium helps to lengthen the columella and vertically lengthens the often small prolabium. The horizontal lip tape is added after the vertical prolabial tape is in place (Fig. 21.7a–e).

Primary surgical closure of the lip and nose is performed from 3 to 5 months of age [10–13].



Fig. 21.7. **a** Bilateral complete cleft with nearly absent columella, wide nasal tip, everted premaxilla and widely separated lips segments. **b** The bilateral nasoalveolar molding plate appliance. Note the horizontal band of soft denture material that joins the left and right lower lobes of the nasal stents. This “prolabial band” spans the columella, pressing in on the nasolabial fold and up at the nostril apices. **c** The bilateral nasoalveolar molding plate in place. Tape is adhered to the prolábium, underneath the horizontal lip tape, and stretches downward to engage the retention arms with elastics. Additional tapes are employed to

support the appliance upward and against the premaxilla and posterior alveolar segments. **d** Following 4 months of NAM therapy, before the primary surgical repair. Note the development of columella. The premaxilla has been gently retracted into the oral cavity and the lip segments are in close approximation at rest. Although not directly observable in this image, the nasal mucosal lining has been stretched by the nasal stents to reduce postsurgical pull down of the projecting nasal tip. **e** Patient at age 1 year, 6 months

21.4 Complications

There are few serious complications associated with nasoalveolar molding. The most common is irritation of the oral mucosal or gingival tissue. Intraoral tissues may ulcerate from pressure or rubbing. Common areas of breakdown are the frenum attachments, the anterior premaxilla, or the posterior fauces as the molding plate is retracted. The infant should be checked at each visit and the molding plate should be properly relieved in all areas that are exerting excessive pres-

sure. The intranasal lining of the nasal tip can become inflamed if too much force is applied by the upper lobe of the nasal stent. Notching along the alar rim can occur if the lower lobe is not positioned or shaped correctly. The area under the horizontal prolábium band can become ulcerated if the band is too tight.

The most common area of tissue irritation is the cheeks. It must be emphasized that the tapes should be removed slowly and carefully so as to avoid skin irritation. The use of tape removal solvents or warm

water can facilitate removal of tapes. If the tissue remains irritated, a skin barrier such as Duoderm or Tegaderm can be used as a base on which the tape-elastic retraction system can be attached. It is sometimes recommended that aloe vera gel be applied to the cheeks when changing tapes.

Poor compliance by the parents can cause loss of valuable treatment time.

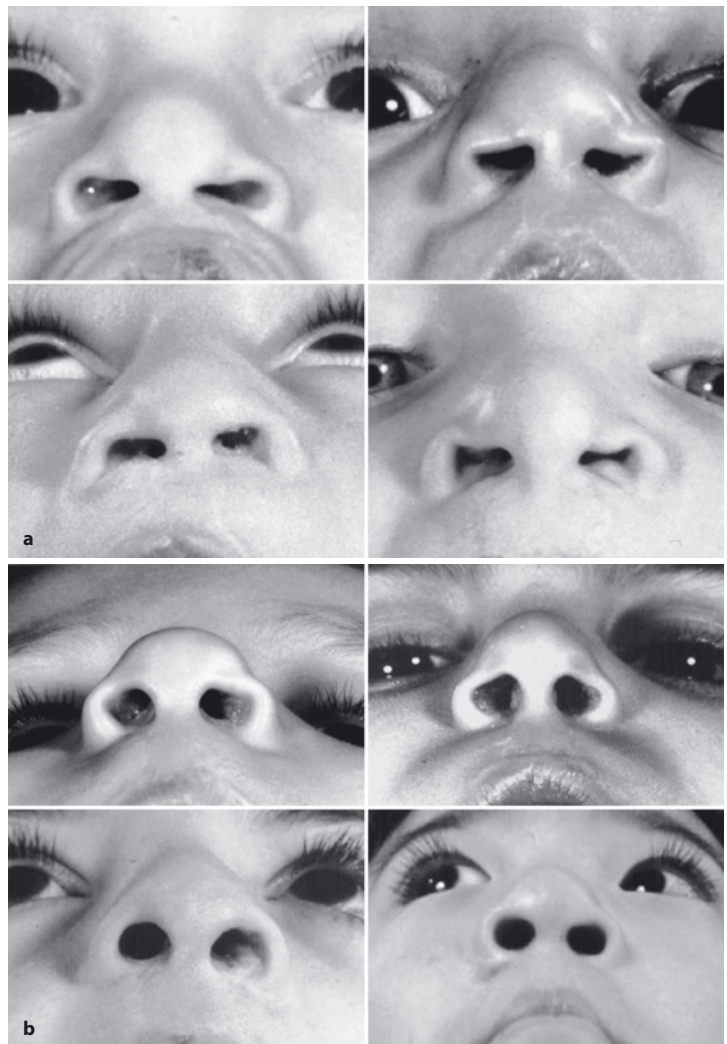
There is a small risk that molding plate will become dislodged and obstruct the airway. Taping the arms too horizontally or with inadequate activation will increase the possibility that the posterior border of the molding plate will drop down onto the tongue. There is only one reported incidence in which this happened, causing a temporary airway obstruction. We placed a 5-mm diameter hole in the center of the molding plate at fabrication to provide for passage of air in the event that the molding plate drops down

from the palate onto the tongue. In the unlikely event that this occurs, the hole, centrally located on the palatal portion of the molding plate, can provide adequate airflow.

21.5 Benefits

The benefits of nasoalveolar molding are numerous. Short-term, the tissues are well aligned prior to primary lip and nose repair, which enables the surgeon to achieve a better and more predictable outcome with less scar tissue formation. Long-term studies indicate that the change in nasal shape is stable [14] with less scar tissue and better lip and nasal form. This improvement reduces the number of surgical revisions for excessive scar tissue, oronasal fistulas, nasal, and labial deformities [22].

Fig. 21.8. a Before the initiation of NAM and the associated surgical technique, there were the inevitable scars at the base of the columella, depressed and broad nasal tips with compromised nasal esthetics. **b** Since the introduction of NAM and presurgical columella elongation, there has been reduced scarring of the lip nose complex, improved nasal tip projection and over all nasolabial esthetics



Overall, fewer surgeries result in substantial cost savings for families and insurance companies [6]. Another important benefit of nasoalveolar molding is the opportunity for the parents to actively take part in the habilitation of their child.

Nasoalveolar molding has evolved over the past decade into its present form through contributions made by practicing clinicians and parents. This method of treatment requires attention to detail with appliance adjustments that are at times less than a millimeter in dimension. Clinical skills in nasoalveolar molding develop with practice, much the way one acquires mastery of the more rigorous techniques in clinical practice. Efficiency in treating patients increases as these clinical skills improve. A significant saving of time may be achieved by the training of a dental assistant or laboratory technician to make adjustments to the molding plate under direct supervision of a skillful clinician. Since the initiation of NAM and the associated changes in the surgical technique which have taken advantage of the NAM procedure, there have been significant improvements in the outcome of primary surgical cleft repair (Fig. 21.8a, b).

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22 Surgical Treatment of Clefts of the Lip and Palate from Birth to Age Ten

S. ANTHONY WOLFE, RAMI GHURANI, MARTA MEJIA

Strategy is the science of planning and directing a large-scale military operation; tactics are the specific maneuvers used to gain advantage over an adversary in a specific encounter.

In this chapter, we will outline the overall strategy that has been used in our unit over the past 30 years in dealing with newborns with clefts of the lip and palate, and we will show a number of patients with a variety of types of clefts, and the specific surgical – or tactical – methods that we used to treat them.

The care of a child with a cleft involves a number of specialists, and in our opinion it is essential that these children be followed by a multidisciplinary team, the members of which all participate in state and national organizations dedicated to the care of individuals with clefts, such as the Florida Cleft Palate Association, and The American Cleft Palate/Craniofacial Association. Our team at the Miami Children's Hospital is headed by a pediatric geneticist; there are several plastic surgeons, orthodontists, pedodontists, otolaryngologists, and speech therapists who regularly attend the team meetings. A clinical psychologist, a social worker, and a nurse coordinator are also regularly present. We often have a parent advocate attend the meeting as well. We feel that this is the *minimum* staffing that a large clinic should have. Other dental specialists such as oral surgeons, prosthodontists, and periodontists should be available to help with specific problems, and in today's medical climate where it is often a battle just to be able to provide necessary care, an attorney and a lobbyist in the state legislature have important contributions to make.

A team is a group that works together with a common goal, although they may have quite different areas of expertise. Understanding each others' disciplines will improve the ability of the team to come to an agreement on how to put together a treatment plan for a specific patient; it is a good idea to have the speech therapist come to the operating room to see a palatal repair or pharyngeal flap, just as it is good for

the plastic surgeon to see how the speech therapist carries out a speech evaluation. However, although consensus is sought for, there are at times conflicting priorities between the different disciplines: a speech therapist might like a palatal cleft closed quite early, in order to avoid the development of compensatory "bad habits," whereas the orthodontist, and sometimes the surgeon as well, would rather close a large cleft at a considerably older age, feeling that there would be less of a chance of causing a growth disturbance or having to deal with a palatal fistula and scarring.

22.1 Timing of Treatment

It is not unusual now to have the diagnosis of a cleft lip made prenatally by the ultrasonographer. This is good, in that the parents can have an opportunity to meet with the surgeon, and perhaps other parents of cleft children, before the birth of their child. This will help avoid the situation that has stuck in so many parents' minds as a very disturbing and unsettling moment: they suddenly learn that their eagerly anticipated baby has a major, disfiguring "birth defect," about which they know very little, and find that the obstetrical and nursing staff don't seem to know very much either. The sooner a member of the Cleft Team, or a member of a Parent Network, can get to the hospital to talk to the new parents, the less these parents will remain in a state of uncertainty and confusion about the future outlook for their child.

We have a Newborn Clinic where parents of a new baby with a cleft can come soon after birth, to get some specific information from the Nurse Coordinator about important issues such as feeding, and if possible to be seen by the surgeon, who will give his or her approach to the particular anatomical problems presented by their baby. A follow-up visit with the surgeon will provide an opportunity to see photographs of similar cleft children operated on by this surgeon

(showing a brochure or textbook with another surgeon's results, although educational, somehow does not seem altogether honest). Initial photographs can be taken, and scheduling made for subsequent treatment.

22.2 Presurgical Treatment

If the cleft involves only the lip, without a cleft of the alveolus or palate, no preliminary treatment is necessary. There are surgeons who advocate early closure of clefts of the lip – as early as 1 week of age [1]. In a good pediatric hospital, this can certainly be done safely. The advocates of this approach state that the scar is better, and that the parents are spared the psychological trauma of seeing their baby with an unrepaired cleft on a daily basis. Both of these statements are true, but we do not think they are sufficient reasons for operating so early. No one has reported on performing alveolar closure or primary nasal correction in the immediate newborn period, and we feel that these procedures should be an integral part of the primary repair. Also, there are limits to the precision that a surgeon can bring to bear on a cleft lip repair, even with the use of 3.5× loupe magnification. In a 6 or 7 pound infant, a 0.5-mm discrepancy would be the same as a 1.5-mm discrepancy in an 18 or 20 pound child. Almost all of the children we have seen who have had their clefts repaired at 1 week of age have had discrepancies that have required that the lip be taken completely apart and done over again. We prefer to repair clefts that involve only the lip at 5–6 months of age, since we have larger structures to deal with, and can have confidence in our primary nasal correction since the alar cartilage is much more mature and free of the softening effects of maternal estrogen.

When there is a cleft of the alveolus and palate as well, we have the luxury of having an expert in naso-alveolar molding (NAM) as part of our team (M. Mejia). For about 25 years the senior author was the associate of Ralph Millard, and did all of the alveolar bone grafting and orthognathic surgery, and much of the palatal surgery that was done in our unit. However, only towards the end of this period, around 1995, were a significant number of primary clefts treated, since before that they were referred to Millard in deference to his tremendous expertise. When we began treating primary clefts in earnest, we used Millard's protocol, which included use of the Latham appliance [2]. This was generally quite successful in bringing the edges of the alveolar cleft into close enough approximation that they could be closed by a gingivoperiosteoplasty (GPP). The apparatus required a general anesthetic to be placed (in our hands at least – Ralph Latham would put them in the office under sedation

and local anesthesia, something we viewed with a bit of trepidation), and would get the job done in 2–4 weeks. There were also some equipment failures, particularly in the bilaterals, that required another anesthetic for correction. One of our orthodontists (Berkowitz) has repeatedly expressed his reservations with the Millard/Latham approach, and has shown that there is a higher incidence of anterior crossbite, and greater difficulties in orthodontic treatment, than with previous patients treated without the Latham/Millard approach and GPP [3]. It has been demonstrated by cephalometric analysis that use of the Latham/Millard approach and GPP resulted in a greater incidence of midface retrusion than patients treated by the older method of leaving the alveolar cleft open until 8 or 9 years old, and then performing alveolar bone grafting along with closure of the alveolar cleft.

The passive approach, or NAM, was initially championed by Court Cutting and Barry Grayson of New York University [4], and when Dr. Mejia arrived with experience in this method, and became part of our team, we were delighted. We no longer had to find a place on our operative schedule to place the Latham appliance, and we had the added benefit of preoperative nasal molding, which aided in the nasal correction, particularly in the bilateral clefts. NAM could be started earlier, at a month or so of age, and generally took 2–3 months to bring the alveolar segments into a good relationship (ideally, a 1 to 1 1/2-mm separation). When the alveolus was in a good relationship by 3 months of age, one could either go ahead with a GPP and a lip adhesion at that time, or keep the appliance in place with occasional adjustments until the definitive lip and nose correction at 6 months of age. From a cost-effectiveness point of view, the latter approach is probably preferable.

Is the GPP inherently a bad thing? That inference has been made, after examining the results of the Millard/Latham approach coupled with the GPP. (see Chap. 20.) At some point, after enough time has passed, we will examine the results of our own series of GPPs, and compare them with those done after using both the Latham appliance and the NAM.

The NYU group has looked at their experience with NAM and GPP in terms of bone formation, and found that 60% had excellent bone formation and would require no subsequent alveolar bone grafting. In the 40% that required subsequent alveolar bone grafting, the results were better than in patients in whom the alveolar cleft had been left open for subsequent closure (perhaps because the soft tissue was almost or completely closed) [5].

Dr. Millard, towards the end of his career, stated that one of his goals would be to be finished with a child with a complete cleft of the lip and palate by age 2 years. We share this goal, although we are not

certain how often it will be reached in our hands. Some patients will require further revisional work on the lip and nose, some will require further alveolar bone grafting in spite of the GPP, some will have VPI even after fastidious palate repair, and some will require maxillary advancements. How often we can really be finished by age 2 can be taken as the benchmark of our success as cleft surgeons.

Although we have adopted the overall strategy for cleft treatment espoused by Millard, there are a few areas in which we have acted differently. Some of these changes he might have adopted himself, and others not.

For instance, in the primary nasal correction, we have used an approach based more on McComb than on Millard. In the McComb approach [6], the alar cartilage on the cleft side is completely separated from the skin through the columellar incision and the alar base incision; a suture is then passed through the desired crus of the lower lateral cartilage through a vestibular approach, without an alar marginal incision. McComb stressed the importance of getting the alar cartilage perfectly corrected at the *beginning* of the operation, before the underlying nostril floor and lip are closed. This made great sense to us, since in secondary nasal corrections performed at 9 or 10 years of age, it was very difficult to get the slumped alar cartilage properly elevated, since it was restrained by scar attachments to the nasal floor that even a very extensive dissection would fail to release.

Millard [7] performed his nasal correction through an alar marginal incision, and through this incision passed a suture between the desired crus of the lower lateral cartilage and suspended the cartilage to the septum. (This was performed after the correction of the lip.) A nasal correction performed in this manner is technically quite difficult in a 6-month-old child, and there is a risk of developing nostril stenosis with the alar marginal incision. Whatever the approach, the essential element in the primary nasal correction seems to be the complete freeing of the nasal skin overlying the lower lateral cartilage, allowing it to be shifted independently of the skin. We have found that the postoperative nasal correction is significantly improved with the use of nasal stents (available from Portex) for 1 or more weeks postoperatively: were these available to Dr. Millard during his career, he would certainly have used them.

Another area in which we have altered the Millard protocol is in the treatment of complete and incomplete bilateral clefts. The Millard approach, which he recommended even for most incomplete bilateral clefts [8], turned down the prolabial vermilion for central vestibular lining, brought the lateral lip vermilion and muscle to the midline to form an “artificial” Cupid’s bow, and banked the skin taken from the

edges of the prolabium to be used as forked flaps for subsequent columellar lengthening. These clefts were treated exactly as if they were complete bilateral clefts. However, even bilateral clefts felt to have a very good result often have an upper lip that is short, and tight relative to the lower lip.

For this reason, incomplete bilateral clefts, either asymmetric or symmetric, we have preferred to deal with as two separate unilateral clefts, and treat them with staged rotation advancements and nasal corrections. This requires two separate operations in order to preserve blood supply to the prolabial skin, and can result in a prolabium which is wider than the published anthropometric norms for noncleft individuals, both of which are disadvantages. However, the preservation of a prolabial dimple with a natural Cupid’s bow, a longer and fuller upper lip, and adequate nasal correction and columellar length, are advantages of this method, and we think outweigh the disadvantages.

For complete bilateral clefts, presurgical NAM has been a godsend for us. The lip is closed as Millard suggested (perhaps someday we will be brave enough to treat them with staged rotation advancements), but the tissue from the sides of the prolabium is not preserved for subsequent forked flaps. The NAM in itself provides a substantial amount of columellar lengthening, and bilateral McComb dissections and alar cartilage suspension, along with nasal stenting, have made it possible with this protocol to avoid using the forked flap.

22.3 Palatal Closure

We completely agree with Berkowitz’s statement that the timing of cleft palate closure should depend upon the size of the cleft – or the cleft space relative to the surface area between the maxillary alveolar ridges [9]. (see Chap. 17) A small cleft involving the soft palate alone can be closed at 1 year of age, or even slightly before. Even in these cases we do a fairly extensive dissection, skeletonizing the greater palatine vessels and separating the levator muscle from the posterior border of the hard palate so that the muscle can be sutured together in the midline with a proper transverse orientation (call this an intravelar veloplasty if you wish).

More complete clefts of moderate width, and involving part or all of the hard palate, are closed at around 18 months of age. In complete clefts of the lip and palate, this is much easier if the alveolus and anterior palate have been previously closed by the GPP. The procedure we generally use involves the development of two posteriorly based mucoperiosteal flaps [10] based on the greater palatine vessels, skele-

tonized with muscle release as mentioned above, and often a more extensive subperiosteal dissection which goes behind the maxillary tuberosity and onto the hamulus (which is not fractured). After the levator muscle is completely dissected from the posterior hard palate, the nasal lining layer is dissected extensively from beneath the maxillary shelves; when available, a medially based flap (or flaps, if a bilateral cleft) is dissected from the vomer. Ideally, the entire nasal lining should be able to be closed with 6-0 Vicryl sutures without tension. The retropositioned levator muscle segments are then sutured together with 5-0 Vicryl, and the remainder of the palatal flaps closed with 6-0 and 5-0 Vicryl. It should be emphasized that the enemy in palatal repair is tension, and when the palatal flaps need to be closed with larger suture material than mentioned above, the likelihood of developing a palatal fistula increases. The way to avoid tension is to wait until palatal growth has provided enough tissue to work with, and to do an extensive dissection of the palatine vessels, releasing the periosteal sleeve [10] and performing an osteotomy of the medial portion of the greater palatine foramen [19] if necessary.

We have on occasion performed the Furlow double-opposing z-plasties [11] as a primary procedure, but this is not any easier in a wide cleft, and results in a longer operation and greater blood loss. The realignment of the levator musculature is probably more physiologic with a two-flap procedure and intravelar veloplasty. We prefer to use the Furlow as a secondary procedure in patients with velopharyngeal insufficiency (VPI) who have a relatively small central gap demonstrated on videofluoroscopy, or in patients with submucosal clefts and VPI. Videofluoroscopy and nasopharyngoscopy will provide information which will help select the appropriate procedure (either pharyngoplasty or posterior pharyngeal flap) in other patients with VPI.

22.4 Secondary Repair of Palatal Defects

If a small palatal fistula occurs after a cleft palate repair, in our hands it usually occurs at the hard palate–soft palate junction. These small defects should be left alone for several years, since many of them will functionally close with further medial palatal growth, and if there is no passage of fluids into the nasal passage, they will not require further surgery. If there is a persistent defect after several years, this can be closed by a secondary procedure. This will be as extensive a procedure as the first operation, if not more so, since careful closure of both the nasal and palatal layers will be necessary. Interposition of a

spacer such as temporal fascia or crushed ear cartilage has improved our results in the closure of small fistulae. For larger fistulae, there may simply not be enough tissue present for a good closure of both the palatal and nasal layers, and on occasion a superiorly based pharyngeal flap may be necessary to provide some further tissue.

For very large palatal defects, local tissues, even with a pharyngeal flap, will not be sufficient to close the “hole.” If the defect is in the anterior portion of the palate, and there is an alveolar defect, a temporalis muscle flap, as espoused by Tessier [12], provides an easy solution. However, if there is an intact alveolar arch, the problem is more difficult. Tongue flaps have been used in the past, and our experience with them has been fairly dismal. It is a miserable thing to put a patient through, and we had a number of failures where the patient either bit through the flap or dislodged it. The FAMM flap [13], which uses vascularized buccal mucosa, has the same drawbacks as the temporalis muscle if there is an intact alveolar arch.

We have found that the microsurgical transfer of a radial forearm flap has provided the surest, most reliable, and easiest-on-the-patient solution to very large palatal defects, whether from poor outcomes with cleft palate repair, or defects from cocaine abuse [14].

22.5 Alveolar Bone Grafting

We have performed alveolar bone grafts on a certain number of patients who were treated by Dr. Millard with an early GPP. Dr. Millard did about 200 of these, and we have bone grafted perhaps 15 – an incidence of 7% or 8% – although there certainly may have been some patients who went elsewhere for treatment, or did not receive necessary treatment. As stated above in the statistics provided by the NYU group, these are very easy alveolar bone grafts, since the soft tissue is either closed completely, or with only minor openings. In contradistinction, a patient with a bilateral cleft and wide alveolar defects communicating with a large defect behind the premaxilla is a formidable challenge, since there is just not enough local tissue available to cover the bone grafts on two sides, even after completely redissecting the palatal flaps; for this reason, many of these bone grafts fail. Cuspid substitution, or moving the entire tooth bearing maxillary segment forward to close the dental gap is a possibility [15], but is also a fairly difficult procedure.

The timing of alveolar bone grafting varies between different teams. Some wait until between 9 and 11 years, just before the eruption of the permanent cuspid. Since approximately 60% of cleft patients will also have a lateral incisor on the cleft side, we feel that earlier bone grafting – at 5 or 6 years – may allow the

lateral incisor to erupt through autogenous bone as well [16]. The orthodontist should have begun a maxillary expansion if required before performing alveolar bone grafting, although the occlusion does not have to be perfectly arranged.

The source of donor bone does not matter, as long as it provides pure, cancellous, autogenous bone. The tibia, the calvarium, and the ilium have all been used; currently we prefer the ilium, since an adequate amount of cancellous bone can usually be obtained percutaneously through a 2- to 3-mm incision.

22.6 Secondary Operations on the Lip and Nose

Once again, if we are successful with the Millard approach of being finished by age 2 years, these secondary operations – by definition an admission of failure – should not be necessary. However, we are perhaps being a bit hard on ourselves and our colleagues to state that any secondary operation is an admission of failure. Perhaps it would be more fair to state that a perfect result has not been obtained – quite a different thing.

Whether the imperfect result is our own, or that of another surgeon, it should be dealt with in the same way. If the upper lip is hopelessly scarred and deformed, or simply very tight relative to a protuberant lower lip, an Abbe flap is the best solution [17]. Any prolabial tissue that is present can be shifted upwards to lengthen the columella. Abbe flaps generally are put off until 7 or 8 years of age simply because the child can better understand what is going on; however, they can be done at younger ages if there are compelling indications. If the lip has more minor problems, lesser solutions are indicated: vermilion excesses can be trimmed, vermilion deficiencies can be treated either by V-Y advancements or dermal or temporal fascia grafts, and minor mismatches of the white roll can be treated by small Z-plasties.

There is an old shibboleth that definitive nasal surgery should be postponed until the midteens, and we see no justification for this. If there is a persistent slump of the alar cartilage, this can be dealt with at any time, even at the time of palate closure, by repeating the McComb procedure. If a child of 7 or 8 has a major nasal deformity, such as a major slump of the nasal tip, this can be dealt with by precise open rhinoplasty techniques which build a normal alar cartilage framework and open the airway with septal resection or repositioning.

We also do not hesitate to address major skeletal malformations, such as Class III malocclusion with major midface deficiencies at earlier ages than we did in the past, where we waited until the cessation of facial skeletal growth. Distraction osteogenesis can easily correct severe problems at an earlier age without the need for bone grafting, and with significantly less relapse [18]. One should not criticize a distraction performed at age 8 or 9 years, even if it needs to be repeated later, if a major dysmorphia which is causing a child psychological distress can be corrected. Other chapters will deal with this in greater detail.

A number of cases will be shown which will illustrate the strategic and tactical approaches that we are using as of spring of 2004; several years hence, we expect that there will be a few differences, and our goal will remain that stated by Millard – try to be finished by age 2 years.

22.7 Case Reports

Case 1. Complete unilateral cleft of the lip with a wide alveolar cleft. Treatment consisted of presurgical maxillofacial orthopedic treatment with a Latham appliance begun at about 2 1/2 months of age (Fig. 22.1). This brought the alveolar segments into good alignment with a gap less than 2 mm. A gingivoperiosteoplasty and lip adhesion were then performed. The definitive lip repair by rotation advancement and nasal correction with the McComb were performed at 6 months of age. The palate was repaired at 18 months of age with a 2-flap palatoplasty and intravelar veloplasty. A 3D CT scan was done at age 3 which shows good occlusion; there may be a slight fissure in the alveolus anteriorly, but it appears intact on posterior view. Dental radiographs will be obtained around age 6 to see if alveolar bone grafting will be required; if not, this child will qualify as a success according to Millard's finish-by-2 goal.

Case 2. Another complete unilateral cleft of the lip and palate, with severe nasal distortion. The alveolar gap was 22 mm. Treatment again consisted of a Latham appliance, GPP and lip adhesion at 3 months, and definitive lip and nose repair at 6 months (Fig. 22.2). Palate repair was performed at 18 months, and the operative photographs show the two flaps (Veau) technique.

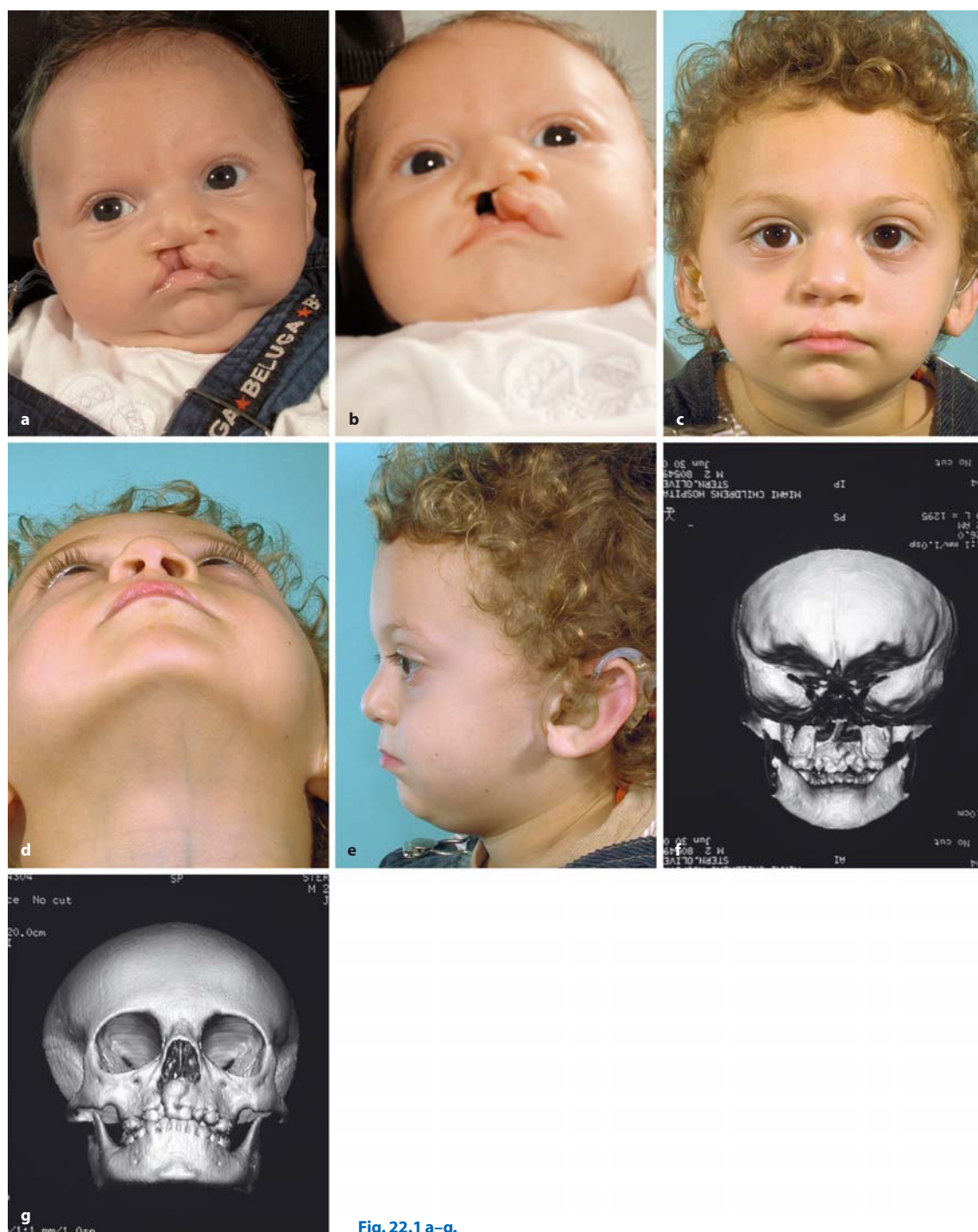


Fig. 22.1 a–g.

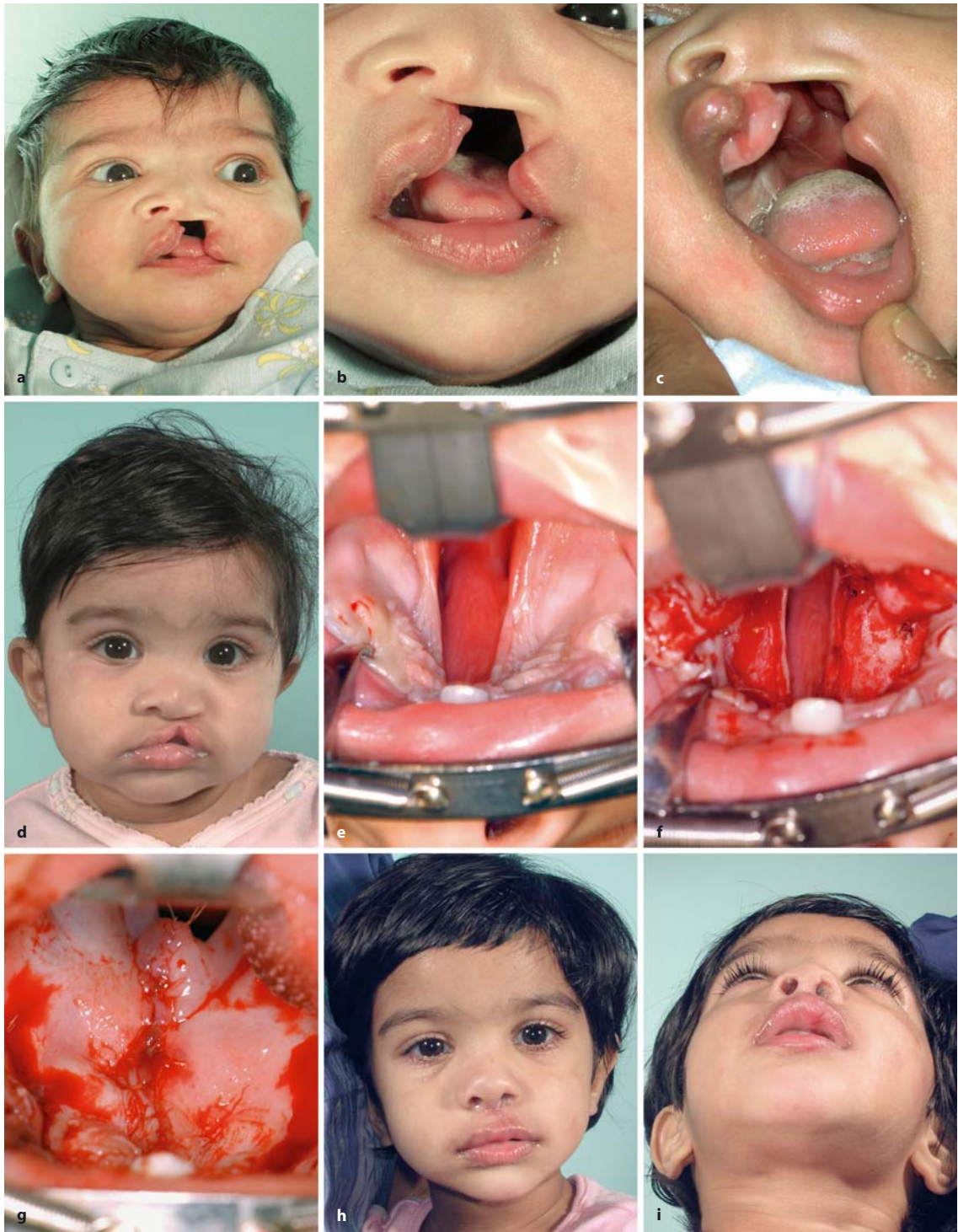


Fig. 22.2 a-i.



Fig. 22.3 a–d.

Case 3. A third complete cleft of lip and palate with a wide alveolar gap, this time treated with NAM rather than the Latham appliance (Fig. 22.3). No lip adhesion

was performed; the definitive lip and nose correction were performed along with the GPP at 6 months of age. Palate closure again at 18 months.

Fig. 22.4 a–d.



Case 4. A child with a wide left complete cleft of the lip and palate, and an incomplete cleft on the right side is shown preoperatively (Fig. 22.4). Presurgical maxillary orthopedics (Latham appliance) were followed by a gingivoperiosteoplasty, and lip and nose

correction, with a rotation advancement and McComb sutures (shown in the operative picture) on the left side at 6 months of age. The right-sided cleft was corrected at 9 months of age with another complete rotation advancement and McComb sutures.



Fig. 22.5 a-f.

Case 5. A complete bilateral cleft and protruding premaxilla is shown preoperatively (Fig. 22.5). Treatment consisted of presurgical maxillary orthopedics (Latham), followed by bilateral GPPs and lip and nose correction at 6 months of age. The columellar lengthening was accomplished by wide dissection of nasal skin from the alar cartilages, removal of intercrural fat, and bilateral McComb sutures. Multiple vestibular effacement sutures were passed, and a nasal stent was maintained for the first postoperative week. The patient is shown at 18 months of age, before closure of the palatal cleft.

Case 6. A very wide complete bilateral cleft of the lip and palate with a projecting premaxilla and very wide alveolar clefts. Initially treated with presurgical maxillary orthopedics (Fig. 22.6) in preparation for the first surgery where the patient underwent a GPP, closure of the alveolar clefts and closure of the anterior palate. One year later, the patient underwent closure of bilateral cleft palate and revision of the lip and nose using the McComb technique, which is shown. The patient is shown postoperatively 2 months after the final procedure.

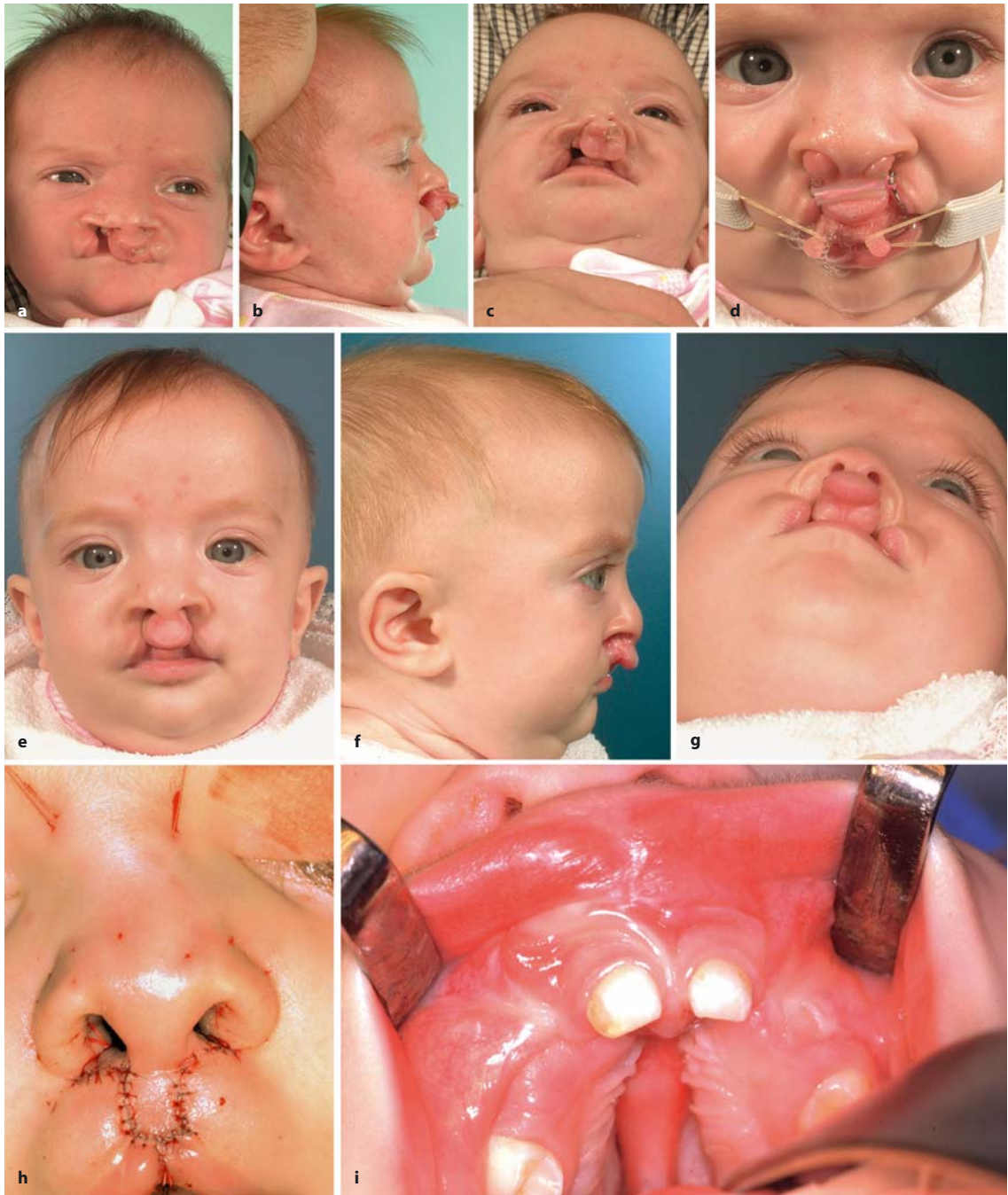


Fig. 22.6 a–k.



Fig. 22.6 a–k. (continued)

Case 7. This patient had primary closure from another surgeon of her incomplete bilateral cleft of the lip using a standard bilateral technique. As the photographs show (Fig. 22.7), she did not have a Cupid's bow, with a fairly tight upper lip and lacking nasal projection. The patient underwent an iliac bone graft to the right alveolar cleft, an Abbe flap, and a cleft lip rhinoplasty which redefined her Cupid's bow, gave her more nasal tip projection, and a fuller upper lip.

Case 8. A bilateral cleft lip was corrected in another country. The preoperative pictures show the patient following a radial forearm flap performed for a very large palatal defect following orthodontic alignment of the premaxilla (Fig. 22.8). The operative pictures show the fabrication of a complete new alar cartilage framework overlying the native alar cartilages, with a columellar strut and spreader grafts (both septal and conchal cartilage was used). There was no reduction of the nasal dorsum. A dermal fat graft was also placed in the central portion of the upper lip. The postoperative pictures were taken at 18 months.

Case 9. This patient had previous repair of a bilateral complete cleft lip by Dr. Millard and had a columellar elongation with a forked-flap. Patient remained with a slumping of the nasal tip, and irregularities of the alar cartilages (Fig. 22.9). Patient underwent a cleft lip rhinoplasty. This improved his tip projection which involved reconstruction and augmentation of the alar cartilages. The patient is shown 7 months postoperatively.

Case 10. This 6-year-old child underwent one previous palatal operation in Cuba, and two subsequent procedures were performed in this country, leading to loss of all palatal tissue from the hard-soft palate junction to the alveolar ridge. A radial forearm flap was performed along with a lip revision, opening the poorly repaired lip completely and thereby avoiding any other cutaneous incision (Fig. 22.10). The procedure was uneventful and the flap had excellent perfusion.



Fig. 22.7 a–g.



Fig. 22.8 a–h.



Fig. 22.9 a–j.



Fig. 22.10 a–g.

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23A Protraction Facial Mask

SAMUEL BERKOWITZ

23A.1 Protraction of the Maxilla Using Orthopedics

Children with complete unilateral and bilateral cleft of the lip and palate are usually at risk for poor facial growth. They are prone to developing midfacial retrusion related to maxillary hypoplasia or growth retardation secondary to excessive palatal scarring. Usually, this results in an anterior dental crossbite or severely rotated maxillary incisors which may occlude in a tip-to-tip relationship with the mandibular incisors. Depending on the age of the patient and the extent of midfacial maldevelopment, some of these early problems can be corrected using midfacial orthopedic protraction forces which increase growth at the circumaxillary sutures as they are repositioned anteriorly (Fig. 23A.1). When all else fails, midfacial surgery is available.

Some of the earlier work in this field, which encouraged a rethinking of the use of orthopedic forces for the correction of midfacial retrusion, includes Hass [1], Delaire [2], Delaire et al. [3–5, 9], Irie and Nakamura [6], Ranta [7], Subtelny [8], Friede and Lennartsson [10], Sarnas and Rune [11], Berkowitz [12], Tindlund [13], Nanda [14], and Molstad and Dahl [15]. More recently this area has been influenced by the work of Tindlund et al. [6–18] and Buschang et al. [19].

Earlier attempts by Kettle and Burnapp [20] in which anteriorly directed extraoral forces were derived from chin caps were relatively unsuccessful. Facial mask therapy seems to offer better control and a wider range of force application.

In many cases, in the mixed dentition, palatal expansion using fixed orthodontic appliances was applied simultaneously with protraction to correct a bilateral crossbite and create a more favorable condition for midfacial growth and development.

Prior to the use of orthopedic forces, many standard orthodontic treatments designed to move the

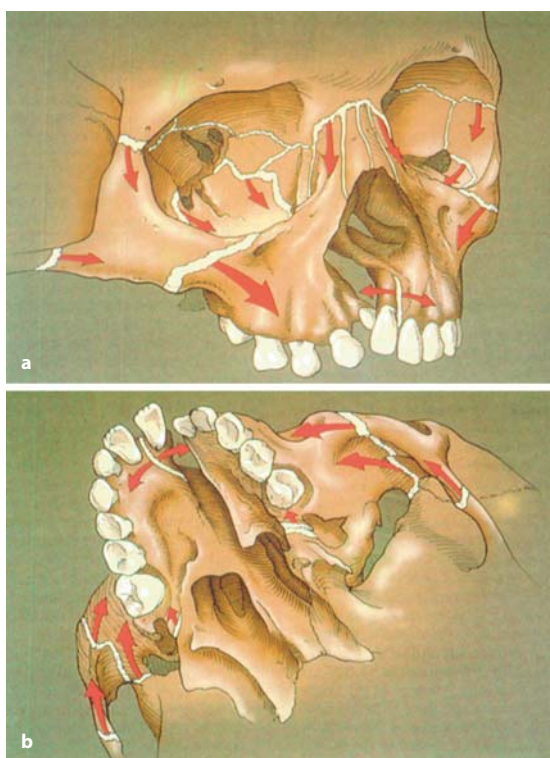


Fig. 23A.1 a, b. Protraction of the maxillary complex using orthopedic forces. The maxilla articulates with nine bones: two of the cranium, the frontal and ethmoid, and seven of the face, viz., the nasal zygomatic, lacrimal, inferior and nasal concha, palatine, vomer and its fellow of the opposite side. Sometimes it articulates with the orbital surface, and sometimes with the lateral pterygoid plate of the sphenoid. Illustration showing how protraction forces applied to the maxilla depend on the disarticulation and growth at all the dependent sutures. (Courtesy of E. Genevoc)

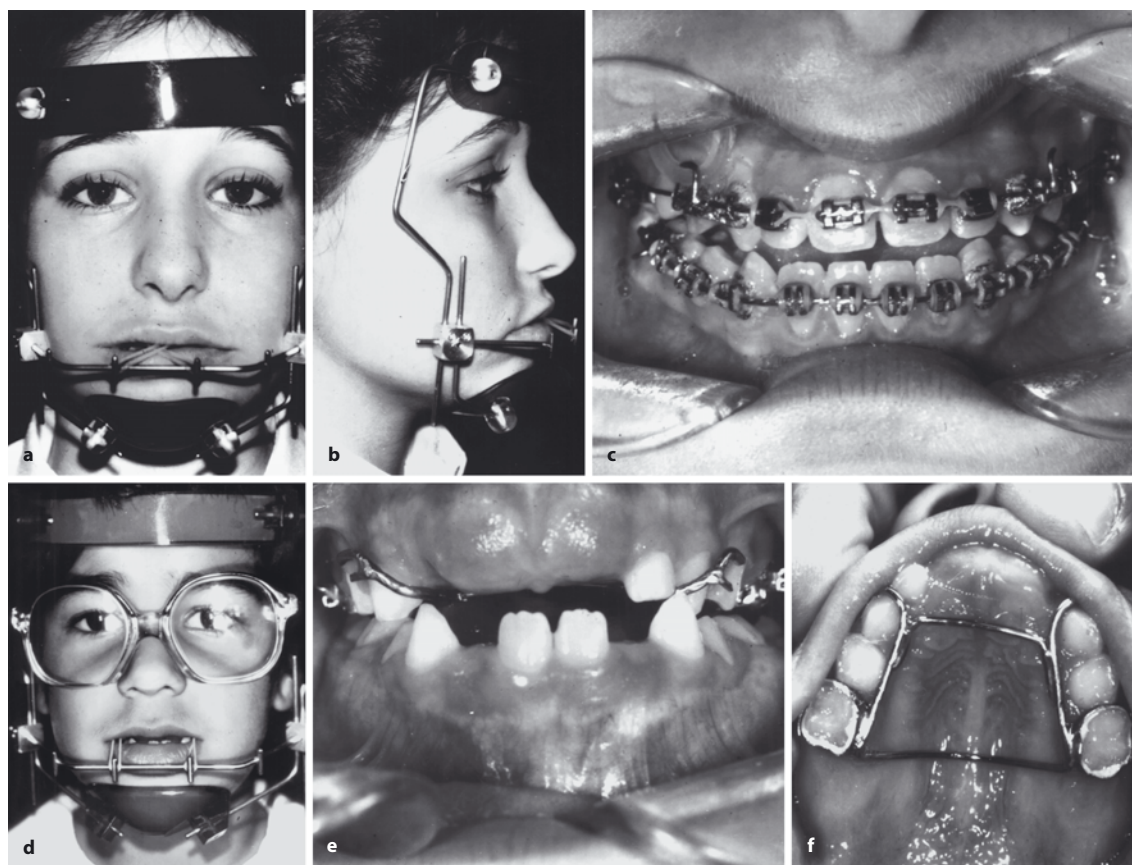


Fig. 23A.2. **a** Frontal and **b** lateral views of a Delaire-style protraction facial mask. Padded chin and forehead rests distribute reaction forces of 350–400 gm per side equally to both areas. Elastics are attached to hooks placed on the arch wire between the cuspids and lateral incisor. **c** Intraoral view of edgewise rec-

tangular arch with hooks for protraction elastics. **d, e, f** Delaire-style protraction facial mask used with a fixed labial-palatal wire framework. Elastic forces of 350–400 gm per side can still be used with this intraoral framework

dentition to correct a Class III malocclusion due to midfacial retrusion in the absence of mandibular prognathism failed. Orthodontic forces applied to the teeth by Class III elastics would not displace the maxilla; at best they would flare the maxillary incisors without creating an adequate incisor overbite and axial inclination. This treatment was found to be unsatisfactory and soon fell out of favor.

Since 1975 Berkowitz has been using a modified protraction facial mask originally popularized by Delaire et al. [3] (Figs. 23A.2–23A.4). It has been very successful in controlling the direction of protruding forces without causing severe sore spots on the chin or forehead. He has found that protraction forces do not modify the direction of mandibular growth as Delaire et al. [3] claimed, but by increasing midfacial height, the mandible is repositioned downward and backward with growth to make the patient's maxillary retrusion appear less evident.

Protraction forces (350–450 gm per side) must be intermittent (the mask is worn only for 12 h per day), and directed downward and forward from a hook located mesial to the maxillary cuspids. Pulling downward from the molars should be avoided because it will tilt the palatal plane downward in the back by extruding the molars and thus opening the bite. When the midfacial height is deficient, protraction forces need to be modified to increase vertical as well as anterior growth. This is done by using more vertically directed elastic forces.

Berkowitz has found 350–450 gm of force per side to be adequate in most instances, but there are rare instances when the elastic force needs to be reduced to prevent sore spots at the chin point. Friede and Lennartsson [10] have used protraction forces between 150 to 500 gm per side. Ire and Nakamura [6] have used 400 gm per side, Roberts and Subtelny [21] 670 gm, Sarnas and Rune [11] 300–800 gm, and Tind-



Fig. 23A.3 a-x. Case BB (WW-62). Maxillary protraction in a UCLP. **a** Complete unilateral cleft lip and palate. **b, c** Lip and nose after surgery. **d** Cuspid crossbite of the lateral cleft segment at 5 years of age due to mesioangular rotation of the

palatal segment. **e** Buccal occlusion after expansion using a quad helix expander. **f, g** 6 years of age. Note relapse of cuspid crossbite due to failure of using a palatal arch retainer. **h** Palatal view showing good arch form

lund et al. [16–18] 350 gm per side. Unfortunately, when performed in the mixed dentition, treatment time may extend into years because of the need to keep pace with mandibular growth. If this is the case, treatment should be divided into intermittent periods not to exceed 6 months at a time with a break for 1 month between periods. Following this formula, the patient will usually remain cooperative.

Although Berkowitz has been successful in using strong elastic forces with labile-lingual appliances during the deciduous dentition, he recommends

starting treatment at 7–8 years of age when all of the maxillary incisors can be bracketed and a rectangular edgewise arch with lingual root torque used as Subtelny [8] suggested. The torqued rectangular arch will carry the incisor roots forward, moving skeletal landmark point “A” anteriorly, which prevents stripping of the alveolar crest with subsequent incisor flaring. The arch wire needs to be tied back so that it does not slide anteriorly, tipping the incisor, rather than moving the entire maxilla forward orthopedically.

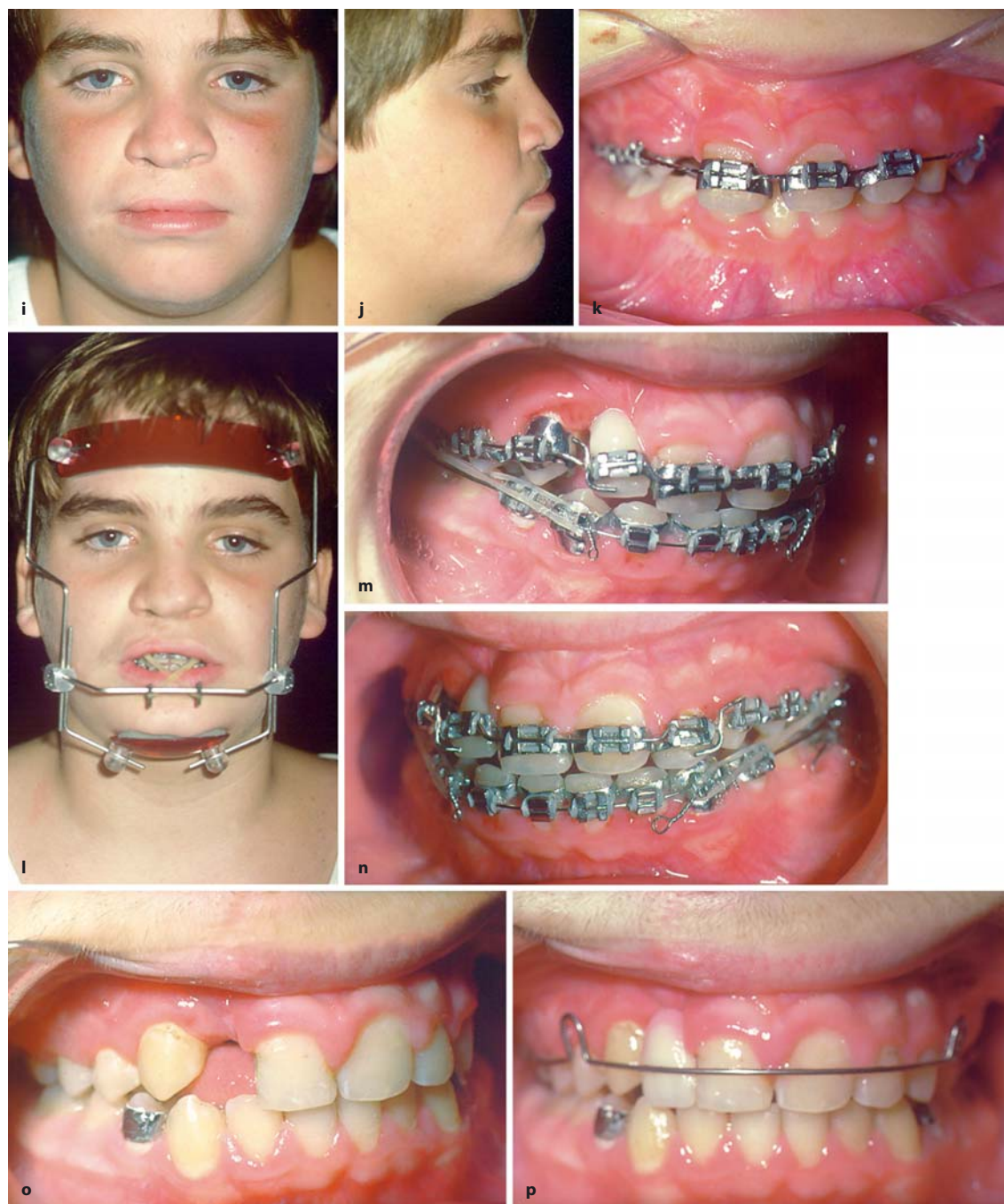


Fig. 23A.3 a-x. (continued) **i, j** Facial photographs at 8 years. **k** Orthodontic alignment of incisors prior to secondary alveolar bone graft. **l** Protraction facial mask with elastics. **m, n** Class III elastics used to maintain tension at circumaxillary suture

during the time not wearing protraction forces. **o** Occlusion after orthopedic-orthodontic forces. Lateral incisor space regained. **p** Removal retainer with lateral incisor pontic

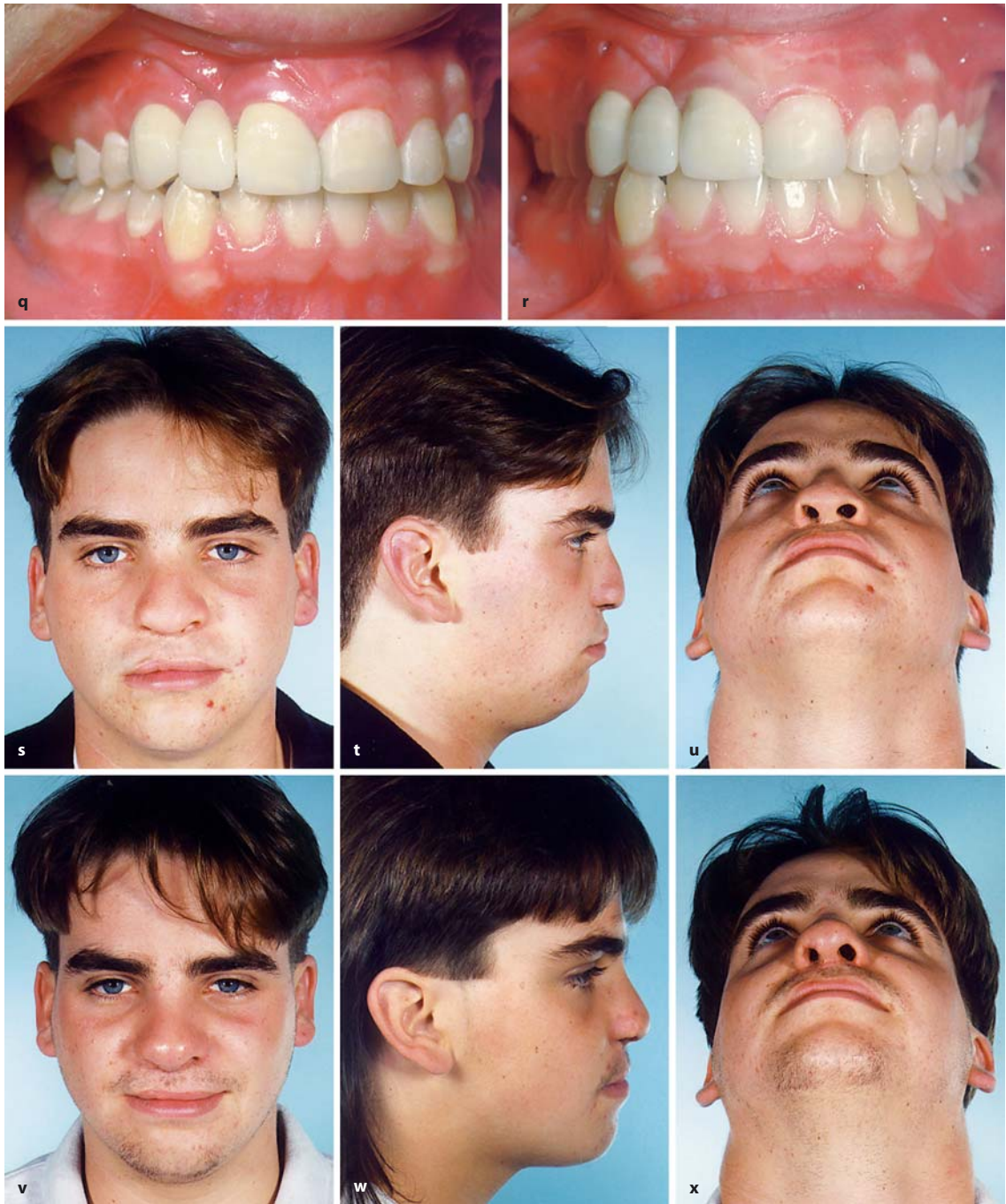


Fig. 23A.3 a–x. (continued) **q, r** Fixed bridge at 18 years of age replacing missing lateral incisor and stabilizing maxillary arch form. **s, t, u** 17 years prior to nose-lip revision. **v, w, x** Facial photos at 19 years, showing good facial symmetry after revision

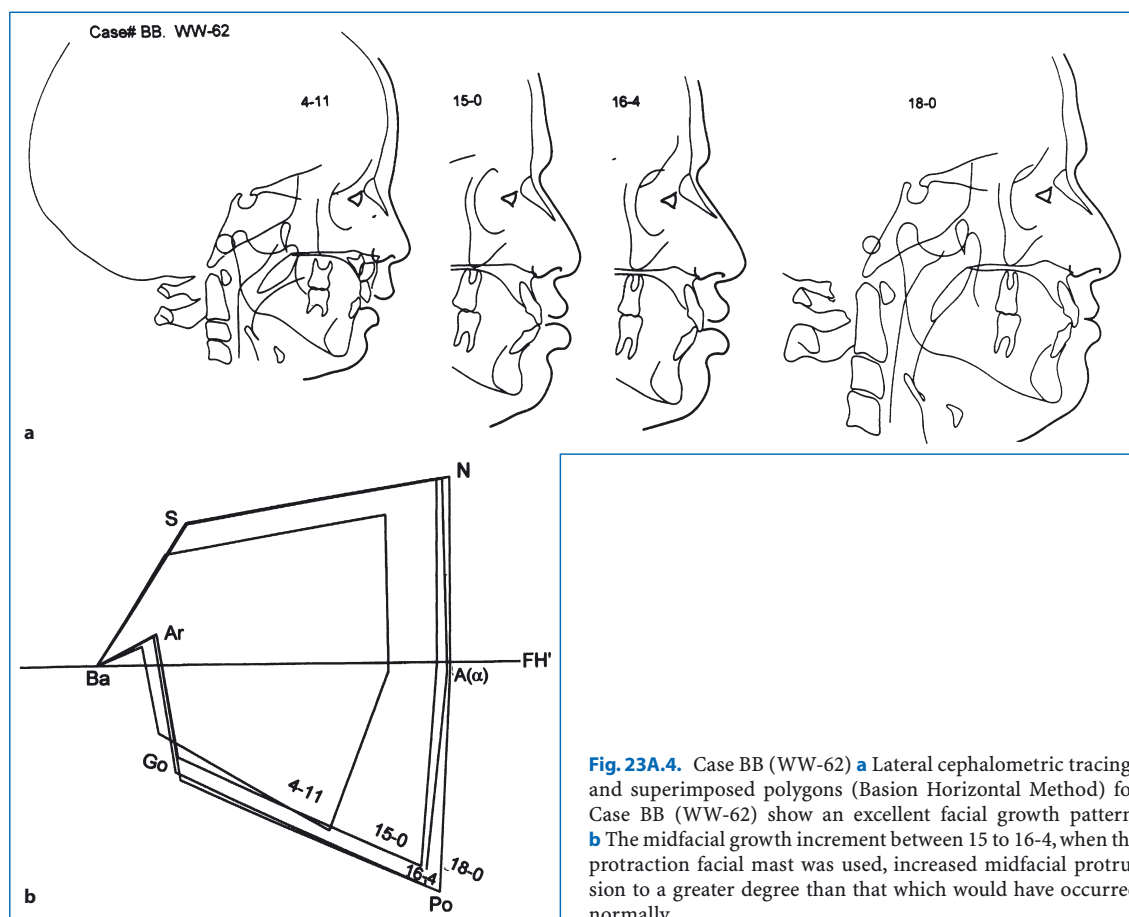


Fig. 23A.4. Case BB (WW-62) **a** Lateral cephalometric tracings and superimposed polygons (Basion Horizontal Method) for Case BB (WW-62) show an excellent facial growth pattern. **b** The midfacial growth increment between 15 to 16-4, when the protraction facial mast was used, increased midfacial protrusion to a greater degree than that which would have occurred normally

Tindlund et al. [16–18] conclude that early transverse expansion of the maxilla together with protraction orthodontic treatment is an effective method for normalizing maxillo-mandibular discrepancies in cleft lip and palate patients. The average age at the start of treatment was 6 years, 11 months, and the average duration of treatment was 13 months. Significant changes were achieved due to anterior movement of the upper jaw and a more posterior positioning of the lower jaw resulting from clockwise mandibular rotation.

Berkowitz also found that the combined use of palatal expansion and protraction forces before the pubertal growth spurt to be a more efficient means of gaining orthopedic advancement than the use of protraction forces alone. He speculates that the expansion forces possibly disarticulate the circumaxillary sutures, thus allowing the maxillary complex to be carried downward and forward more easily.

Delaire et al. [5] and Subtelný [8] have stated that orthopedic forces applied to the entire maxillary com-

plex are more likely to be effective in younger children.

Berkowitz's clinical experience supports the recommendation by Abyholm et al. [22] and Bergland et al. [23] (1) that a rigid fixation of the advanced maxilla should be maintained for at least 3 months after bone grafting, and (2) the use of protraction forces. This is necessary to help reduce the tendency to relapse created by the surrounding soft tissue of the lip, muscles, and skin.

Many patients with a complete bilateral cleft lip and palate have a protruding premaxilla until 10 years of age or older, but after the postnatal mandibular growth spurt, the maxillary incisor teeth may be in crossbite. Protraction orthopedic forces with anterior criss-cross elastics upright and reposition the premaxilla forward, perhaps by inducing bone growth at the premaxillary-vomerine suture. Fixed retention is always necessary to control the improved incisal overbite-overjet relationship at least until secondary alveolar bone grafting is done.

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23B Protraction Facial Mask for the Correction of Midfacial Retrusion: The Bergen Rationale

ROLF S. TINDLUND

23B.1 Early Rehabilitation

Optimal rehabilitation of a child with cleft lip and palate (CLP) involves the achievement of ideal speech, facial aesthetics, and dental occlusion. Dentofacial appearance is of major importance for the development of a child's self-esteem [1–3]. Early adolescence is a time of change and uncertainty and a period of special importance because negative self-esteem developed in these years is likely to be retained into adulthood [4, 5]. Therefore, early rehabilitation is of major importance.

Obtaining an optimal treatment result in complete clefts of the lip and palate is dependent on the prevailing treatment philosophy, clinical skills, and the interaction of the Cleft Lip and Palate (CLP)/Craniofacial Team. The orthodontist is mainly concerned with the achievement of normal long-term facial growth and development, based on his or her ability to recognize, prevent, and treat dentofacial anomalies.

Quality assurance and the cost-effectiveness ratio are important factors that need to be considered in the systematic delivery of health care. Quality assurance focuses on the achievement of the goals and the quality of overall team management based on the usage of accepted physiological principles. Treatment results are not always predictable because patients differ in their facial growth patterns and the nature of the palatal defect, requiring individualized orthodontic treatment plans depending on the developing malocclusion. This philosophy is at variance with the generally held orthodontic strategy, which is to postpone all orthodontic intervention until the permanent dentition [6]. The relative low cost of utilizing interceptive orthopedics at an early age, due to the need for infrequent visits with uncomplicated mechanics, is a reasonable option for the early improvement of dentofacial appearance. An additional bonus to performing treatment at this period is that patients develop a pos-

itive attitude toward themselves and parents to their child's future status.

The specific aim of this chapter is to present a CLP treatment program that incorporates interceptive orthopedics in faces with midfacial retrusion and demonstrate how a fixed orthopedic-orthodontic appliance system may be used for both transverse widening as well as the protraction of the maxilla. Interceptive orthopedics is discussed with respect to treatment timing and anticipated clinical results, reviewing the limitations, and criteria necessary in case selection to improve long-term prognosis.

23B.2 Midfacial Retrusion in CLP Patients

Irrespective of the method used in primary cleft repair and the surgical skill of the operator, a certain number of patients will show an unfavorable growth pattern. Even if one plastic surgeon performs all surgery utilizing the same procedures, and the same treatment protocol, individual outcomes may vary from excellent to unsatisfactory. The variable results reflect individual differences in craniofacial type and growth patterns on which the cleft maxilla is superimposed. Also, one needs to consider acquired variables, such as the degree of prenatal maxillary hypoplasia and facial asymmetry in cleft embryo-pathogenesis and detrimental growth deviations related to the surgical procedure and skill of the surgeon.

Midfacial retrusion may be due to underdevelopment and/or relative posterior positioning of the upper jaw to the mandible. The maxillary growth deficiency usually is three-dimensional, resulting in a shortening of maxillary length and a decrease in width and height. Midfacial retrusion is more often seen in unilateral cleft lip and palate (UCLP) patients [7–11] whereas in bilateral cleft lip and palate (BCLP)

the initially prominent premaxilla become less protrusive over time, achieving an almost ideal incisor overjet–overbite relationship by the late teenage years [12].

Patients with an underdeveloped maxilla, which results in skeletal and/or dentoalveolar discrepancies, often show anterior and/or posterior crossbites with a concave soft-tissue profile. Since 1977 the treatment protocol of Bergen Cleft Palate–Craniofacial Center has included an interceptive orthopedic treatment phase designed to correct anterior and posterior crossbites during the deciduous and early mixed dentition and to obtain optimal alveolar cleft space to enhance tooth eruption and alveolar development. This would ultimately lead to a favorable functional dental occlusion and create better conditions for attaining normal midfacial growth and development [9–11, 13–19].

23B.2.1 Anterior Crossbite

Anterior crossbite (incidence about 3%–5% in Scandinavia) may be found in all facial types – prognathic, orthognathic, and retrognathic – in combination with varying degrees of hypo- or hyperplasia of the jaws. Different sagittal skeletal jaw configurations, some with deep or skeletal open bite may be associated with excessive dentoalveolar mandibular proclination or maxillary retroclination along with the lack of sufficient dental space in the upper arch. Guyer et al. [20] found skeletal maxillary retrusion in two thirds of noncleft Class III children. This is of great therapeutic interest since orthopedic influence seems to be more effective in influencing the sutures of the maxillary complex than in restraining mandibular growth [9–11, 14–17, 21–25]. However, the long-term differential diagnosis between mandibular excess and maxillary retrusion is difficult to determine before puberty [20, 26–30]. For this reason, children with the appearance of midfacial retrusion and anterior crossbite may benefit from an early interceptive orthopedic treatment phase. The need for orthognathic surgery is usually determined after puberty, taking facial appearance as well as dental occlusion into consideration. A family anamnesis of anterior and posterior crossbite is of particular interest in the CLP population because maxillary hypoplasia is a common finding in these patients.

23B.2.2 Orofacial Function

Optimal orofacial function with adequate incisor relationship in the primary dentition are important determinants for normal growth and development of the

anterior part of the maxilla. There is a generally accepted belief that form and function are mutually dependent. This interaction in facial clefts is important because malfunction has been shown to negatively influence facial growth. Thus, the facial characteristics of a noncleft child who is a mouth-breather may show some similarities in appearance to the typical CLP patient [9, 10]. In CLP, midfacial retrusion due to deficient midfacial growth may be aggravated by increased nasal airway resistance, low and forward posture of the tongue, and lack of sufficient stimuli from proper masticatory forces. Early widening of the upper jaw enhances nasal respiration [31–34], while permitting the tongue to assume a more normal elevated position within the mouth [35]. Direction of eruption and the final position of teeth are closely associated with the development of the alveolar process, which in turn is dependent upon the number, size, and location of teeth [36–38]. Early orthopedic treatment which includes transverse expansion and anterior protraction of the maxillary complex will improve the dimensions of the nasal as well as the intraoral space, permitting the tongue to elevate and assume a normal posture within the vault space, thus breaking the vicious circle of poor function leading to poor form with growth.

23B.3 Principles of Orthopedic/Orthodontic Treatment in CLP Patients

The orthopedic/orthodontic CLP treatment protocol in Bergen utilized since 1977 is based on selective periods of active, controlled, efficient treatment followed by intervals of fixed retention, as recommended by American Cleft Palate–Craniofacial Association in 1993 [39]. The easily obtained acceptance of the need for patient cooperation along with an excellent cost/effectiveness assessment ratio support the use of this philosophy of treatment. The following orthodontic treatment phases should be considered as viable options for the individual patient:

1. Presurgical maxillary orthopedics (0–3 months, used in a few cases only)
2. Interceptive orthopedics (6–7 years, about 20% of cleft patients) which involves transverse expansion and protraction (Facial Mask)
3. Alignment of maxillary incisors prior to secondary alveolar bone grafting
4. Secondary alveolar bone grafting of the cleft alveolar process
5. Conventional orthodontics in the permanent dentition is always necessary
6. Dental adjustments dependent on prosthodontic or orthognathic surgery needs (17–19 years)

Individualizing the timing and sequencing of treatment is essential due to the wide range of skeletal malformation associated with dental malocclusions. It is of utmost importance to individualize each treatment plan and to revise this plan at different ages of dental and skeletal development, all of which is conveniently based on a diagnosis-related checklist.

23B.3.1 Checklist for CLP Orthopedic/Orthodontic Treatment Objectives

23B.3.1.1 Presurgical Orthopedics

The plastic surgeon seeks to obtain optimal function and appearance and avoid the need for extensive revisionary surgery by using proven surgical techniques that result in a minimum of scarring and palatal growth impairment. In some cases, presurgical orthopedics can help the plastic surgeon unite anatomical structures with a minimum of force and stress to the tissue. Individual decisions are made by the plastic surgeon.

- Reposition severely displaced maxillary segments.
- Reduce width of very wide clefts.
- Improve symmetry of nose and upper jaw.
(Only used in extreme cases, and in some treatment philosophies this stage is not necessary.)

23B.3.1.2 Interceptive Orthopedics

Transverse expansion followed by anterior protraction of the upper jaw should only be utilized in cases with anterior and/or posterior crossbite with mid-facial retrusion. Treatment should be instituted early enough to allow the permanent incisors to erupt spontaneously into a normal overjet and overbite occlusion (Fig. 23B.1).

- Eliminate anterior crossbite
- Eliminate posterior crossbite
- Create optimal space to permit spontaneous eruption of the incisors
- Improve nasal respiration
- Improve tongue placement

23B.3.1.3 Alignment of Maxillary Incisors

In spite of achieving optimal dental space after transverse expansion, the permanent incisors often erupt rotated and retruded, tipped, or retroclined, placing

them in crossbite. After transverse expansion, alignment of the permanent incisors is easily performed, giving the child a nice dental smile equal to that of his or her classmates (Fig. 23B.2; in Fig. 23B.1 incisor alignment was not needed)

- Straightening of malpositioned incisors
- Creating an optimal aesthetic incisor relationship to the facial midline

23B.3.1.4 Secondary Alveolar Bone Grafting

The use of primary periosteoplasty at age 3 months was rejected after introduction of secondary bone grafting [40]. It is usually performed between 8 and 11 years of age with the orthodontist selecting the appropriate age.

- Eliminate remaining bony clefts and improve bony support of contiguous teeth
- Enhance orthodontic closure of the missing incisor space in the cleft area
- Stabilize of separated jaw segments
- Close oronasal fistulas
- Provide bony support to alar base in cases with nasal asymmetry
- Eliminating mucosal recesses

23B.3.1.5 Conventional Orthodontics in the Permanent Dentition

The orthodontic treatment goals are similar to the general orthodontic principles utilized for noncleft patients: To establish ideal dental function, facial aesthetics and speech. Extraction of mandibular teeth to compensate for a hypoplastic upper jaw is usually not indicated until after the critical mandibular growth period has passed. In CLP patients, a bonded palatal fixed retainer is often necessary after treatment involving arch expansion to avoid relapse of the corrected palatal arch form.

- Improve the relationship of the lips
- Achieve harmonious balance of the dentition in the opposing jaws
- Achieve favorable skeletal maxillomandibular jaw relationship
- Achieve normal incisor overjet and overbite
- Correct dental axial inclinations
- Avoid the use of artificial teeth
- Achieve functional dental occlusion
- Achieve optimal nasal breathing

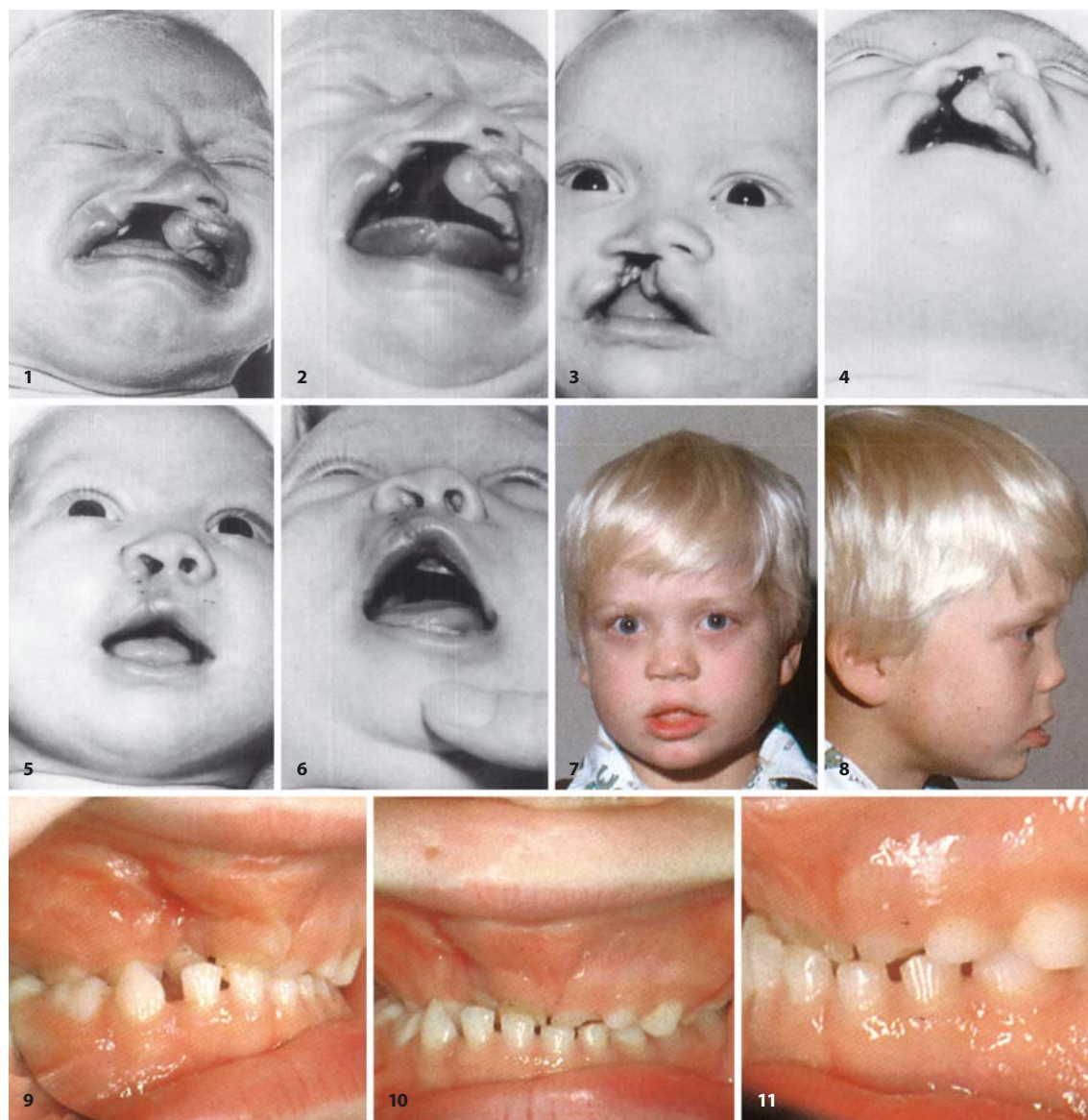


Fig. 23B.1. Complete UCLP, category 2A. (1–2) At birth, January 1975, (3–4) after presurgical orthopedics; (5–6) lip closure at age 3 months; (7–12) at 6 years moderate anterior and unilateral posterior crossbites with a slight concave profile; (13–27) interceptive orthopedics from age 6 years includes transverse expansion for 3 months using a quad-helix, (14) followed by protraction for 6 months using a facial mask, (17–18) and retention using a fixed palatal archwire (15) to encourage spontaneous eruption of upper permanent incisors into normal position. A nice dental smile was achieved without early ortho-

dontic alignment of the upper incisors; (28–33). Alveolar bone grafting at 10.5 years. Two right upper lateral permanent incisors erupted into the cleft area; (34) Facial profile at 12 years (35–41); conventional orthodontics at 13.5 years lasting for 18 months. The two upper second bicusps were missing and the supernumerary right upper lateral permanent incisor was removed; (42–48) dental occlusion at 18.5 years; (49–50) cephalometric graphic analysis at 6, after interceptive orthopedics, and at 15, and 18 years; (51–53) facial appearance at 15 years; (54–59) facial appearance at 18.5 years

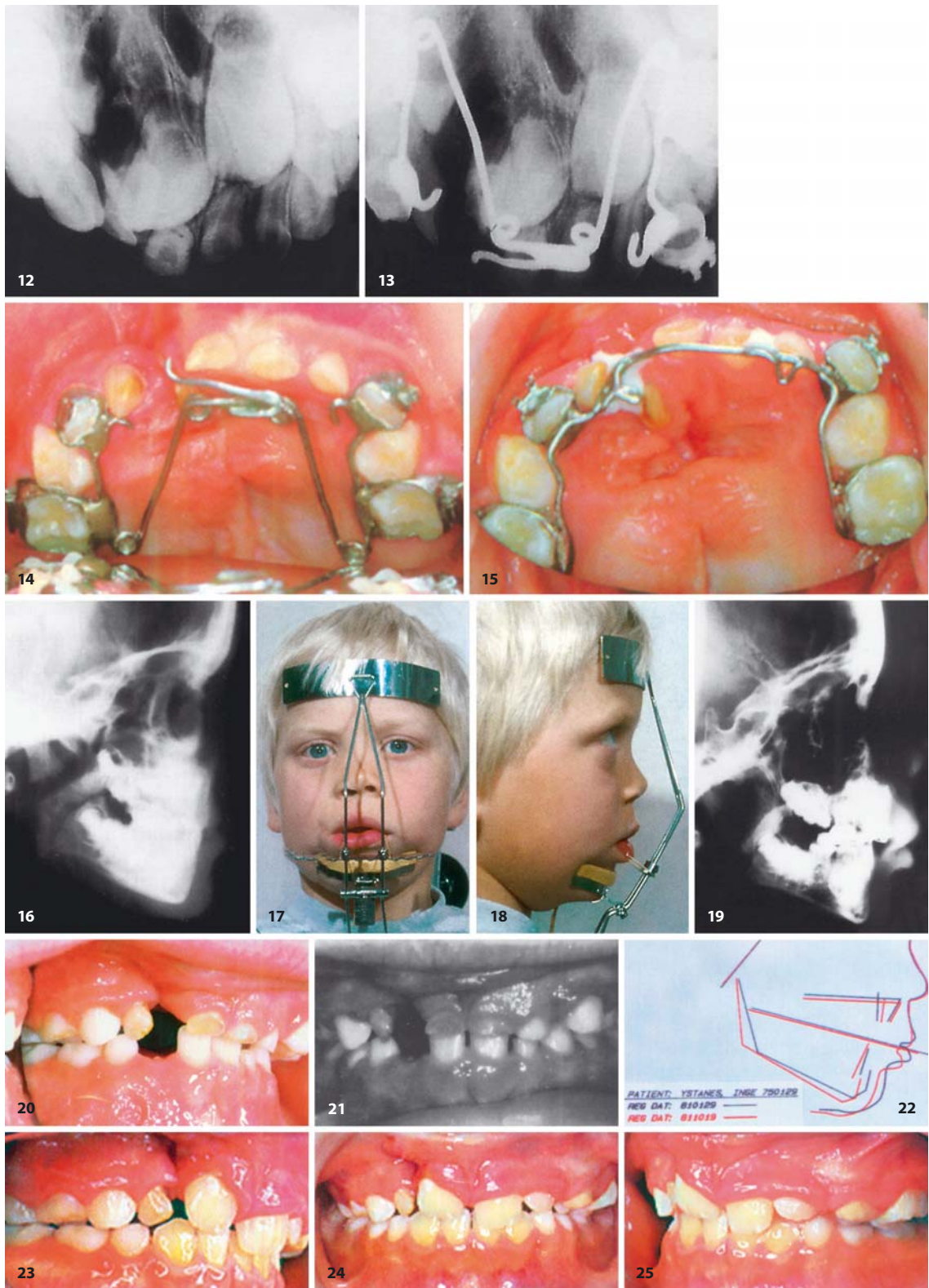


Fig. 23B.1. (continued)

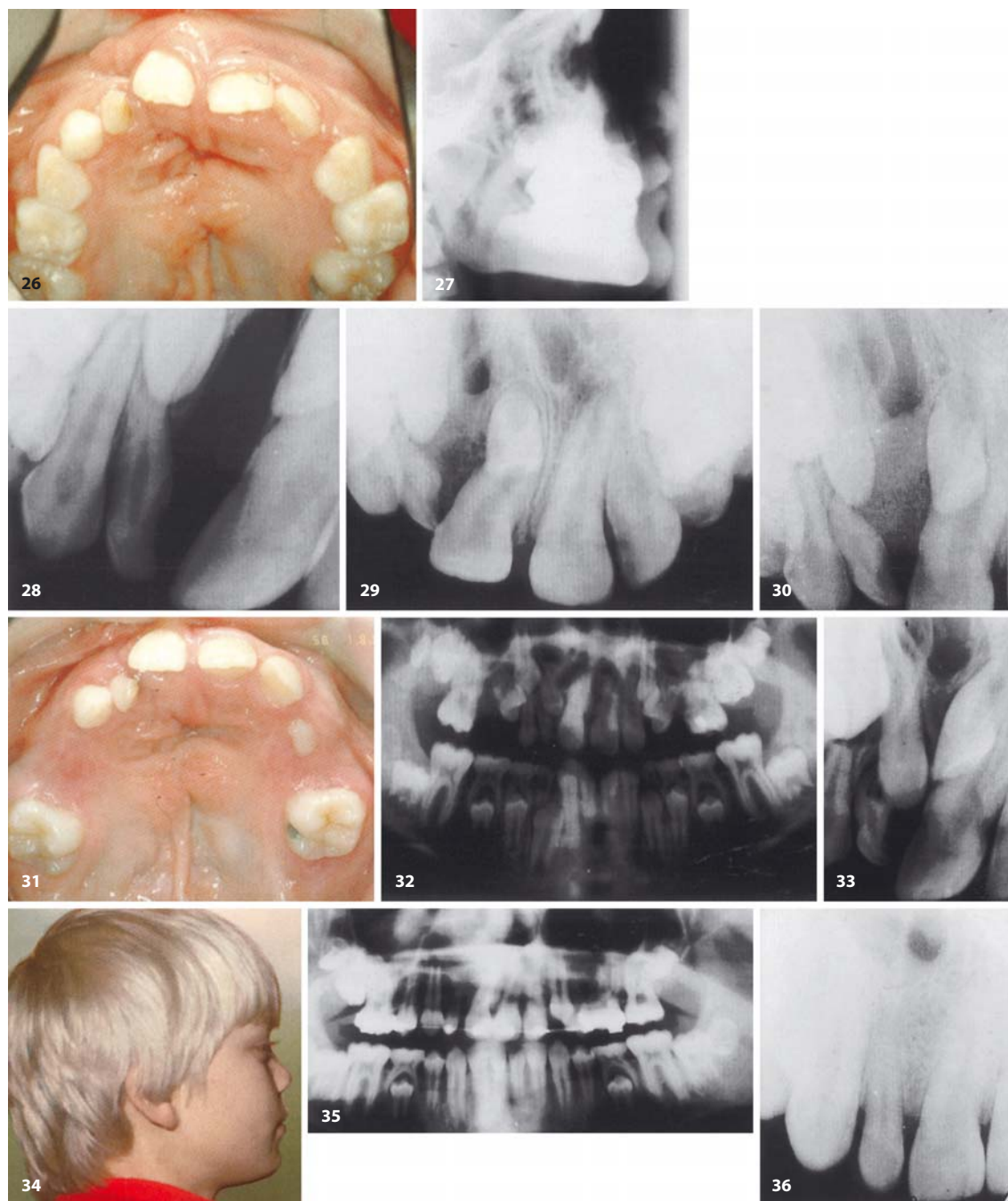


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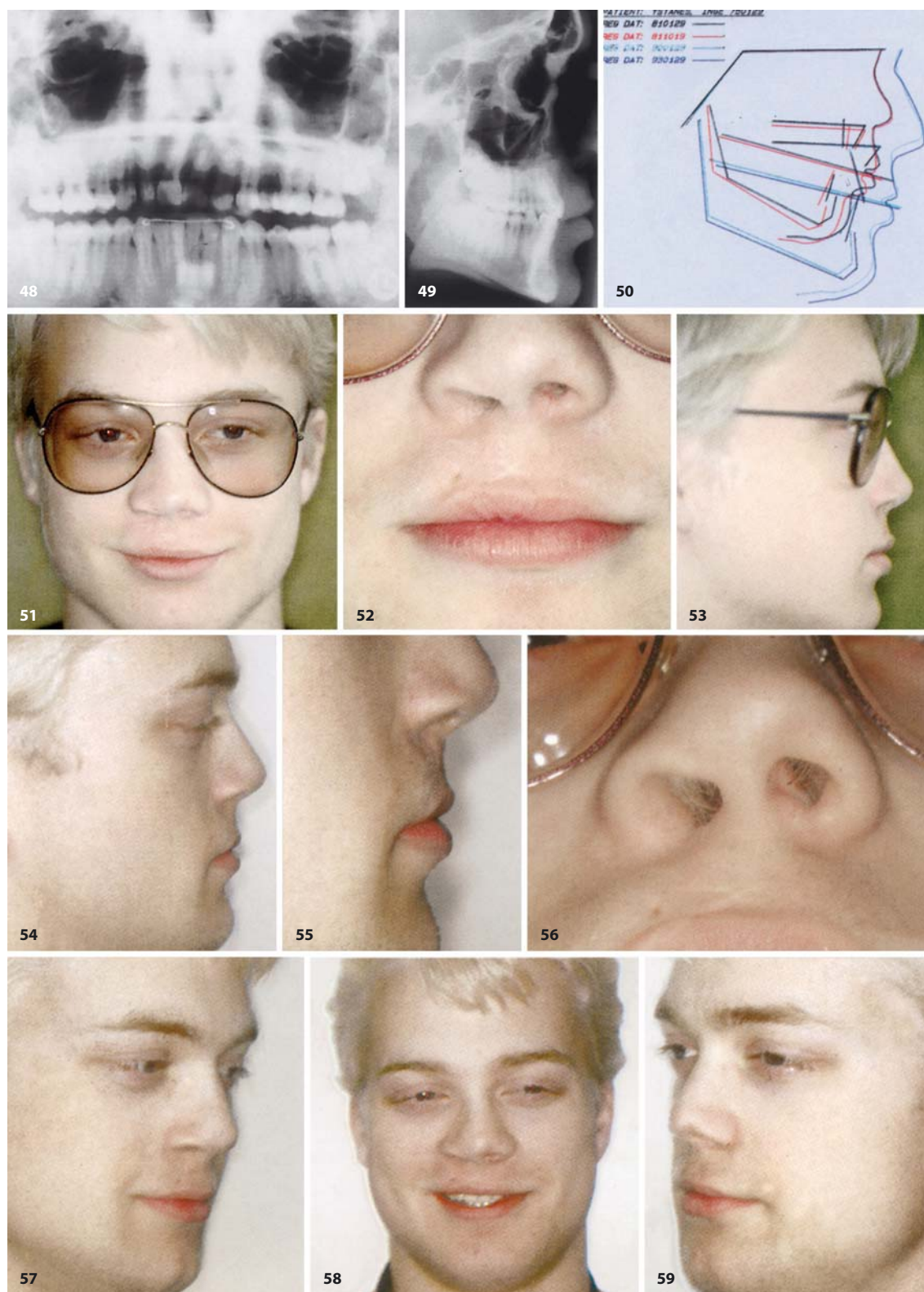


Fig. 23B.1. (continued)

23B.3.1.6 Dental Adjustments at Age 16–17 for Girls, 18–19 years for Boys

In cases with major skeletal jaw discrepancies, orthognathic surgery may be needed to normalize the skeletal jaw relationship and achieve a well-balanced facial appearance with stable dental occlusion. If two or more teeth are absent in the same dental segment, a small bridge is normally needed. However, dental implants are likely to become an important aspect of future prosthetic replacements.

23B.4 Outline of CLP Treatment Procedures in Bergen

To appreciate our treatment philosophy, a brief summary of the treatment approach and concepts of the Bergen Cleft Palate Center will be presented. Along with the Oslo CP Center, it serves a population of 5 million. Due to demographic distribution, many patients must travel distances up to 2,000 km to either center. Hardships are compounded by the need to travel in very cold weather during winter; therefore, the planning and coordination of health services are crucial for optimal utilization of available resources. Treatment costs and travel expenditures are covered by the government's social security program. The Bergen CLP Team treats about 55 newborn babies yearly. Treatment procedures are coordinated between the Department of Plastic and Reconstructive Surgery, University Hospital of Bergen; the CLP Center at the Department of Orthodontics and Facial Orthopedics, Faculty of Dentistry, University of Bergen; and the Eikelund Center for Speech Pathology.

23B.4.1 Plastic Surgery

Since 1986, in complete clefts of the lip and palate, a Millard lip closure is performed at 3 months combined with a single-layer vomerplasty for closure of the anterior part of the palate. The soft palate and isolated palatal clefts are closed at 12 months using a von Langenbeck technique. Alveolar bone clefts are left open until secondary bone grafting at 8–11 years of age. Between 1971 and 1986, the lip closure was combined with a periosteoplasty of the cleft alveolar process [41].

23B.4.2 Interceptive Orthopedics

23B.4.2.1 Protraction Facial Mask

Extraoral heavy forces from a facial mask directed forward and downward from the maxillary cuspid area have been shown to correct midfacial retrusion at an early age [9–11, 14–17, 18, 19, 22, 23]. Protraction from the maxillary cuspid area produces an adequate horizontal and vertical force to increase midfacial vertical height as well as anteroposterior length. In some instances, it also can reduce an anterior open bite by lowering the palatal plane. For this reason, early correction of anterior and/or posterior crossbites during the deciduous and mixed dentition is highly recommended:

Bergen Rationale:

1. Transverse expansion coupled with
2. Protraction of the upper jaw and
3. The use of fixed palatal arch retention after treatment

When considering a treatment plan for young children who travel great distances, it is important to consider patient comfort as well as treatment efficiency.

In cases of marked midfacial retrusion, interceptive orthopedics is started at 6 years and often lasts for 15 months with an average of six visits (two visits for a transverse expansion of about 10 mm during a 3-month period, and an additional four visits for the use of protraction forces for 12 months).

23B.4.2.2 Quad-helix Spring (with Four Bands and Hooks)

A fixed palatal expansion appliance can be easily combined with the use of an extraoral facial mask (Figs. 23B.1, B.2) [13] providing:

1. Controlled transverse expansion when needed
2. Adequate fixation for anterior protraction by a facial mask
3. Use with edgewise appliance for the alignment of incisors
4. Well tolerated by small children without sedation, causing a minimum of discomfort
5. Minimum of chair time
6. Can be easily kept clean

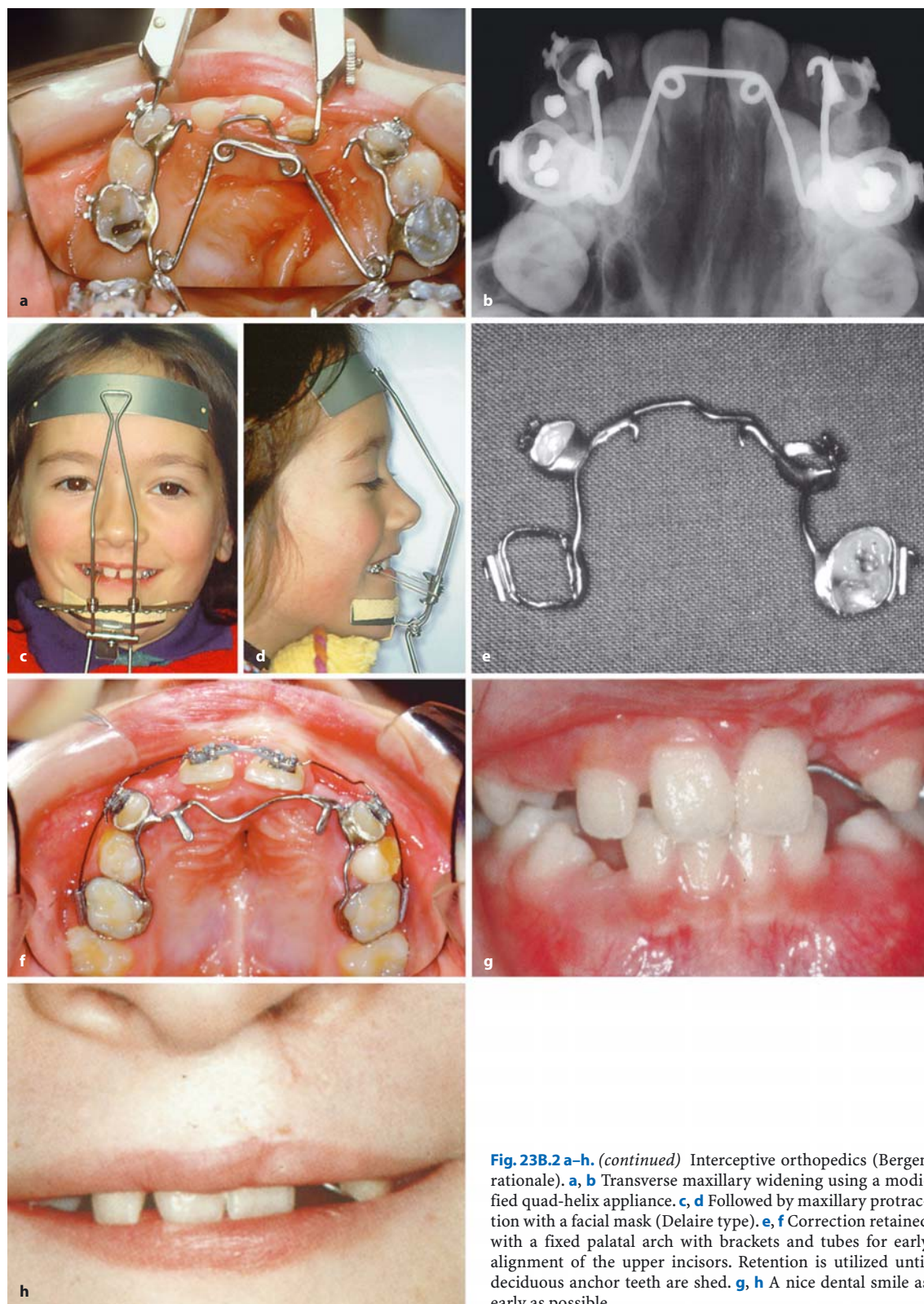


Fig. 23B.2 a-h. (continued) Interceptive orthopedics (Bergen rationale). **a, b** Transverse maxillary widening using a modified quad-helix appliance. **c, d** Followed by maxillary protraction with a facial mask (Delaire type). **e, f** Correction retained with a fixed palatal arch with brackets and tubes for early alignment of the upper incisors. Retention is utilized until deciduous anchor teeth are shed. **g, h** A nice dental smile as early as possible

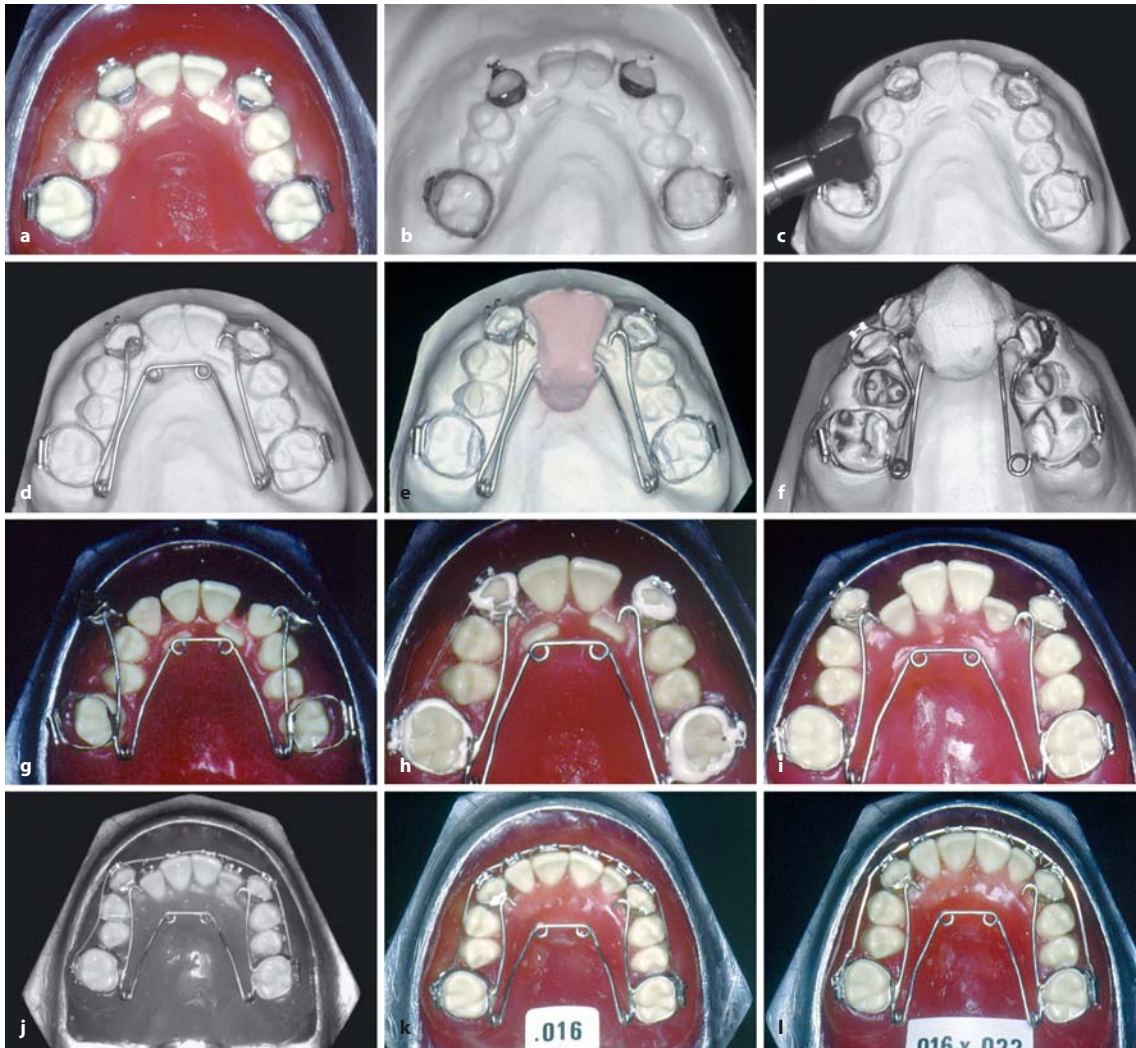


Fig. 23B.3 a–l. Fabricating a modified quad-helix appliance. **a, b** Bilateral posterior crossbite with lack of space for erupting lateral permanent incisors. Bands with brackets or tubes are fitted to the upper deciduous cuspids and deciduous second molars and carefully replaced into an alginate impression. **c** Plaster

removed underneath soldering zones. **d** Quad-helix arms are precisely adjusted. **e, f** Quad-helix arms are soldered to all four bands. **g** Each arm is individually activated. **h** Cemented. **i, j, k** Combined with round labial arches for alignment of incisors. **l** Labial incisor root-torque with rectangular archwire

The creation and use of a modified quad-helix [Rocky Mountain: maxillary quad-helix. 0.38 (0.985 mm) Blue Elgiloy (Ricketts)] appliance [13] is shown in Fig. 23B.3. Four preformed bands with brackets or tubes are placed on the second deciduous molars and deciduous cuspids and are soldered to the quad-helix spring. The first deciduous molars serve as additional anchorage. Permanent molars are banded only when the second deciduous molars are missing. Hooks for elastics are positioned mesio-lingually to the cuspid bands. The elastics are attached to the protraction facial mask (Fig. 23B.2c, d). The elastic forces are direct-

ed forward and downward from the anterior maxillary segment, resisting the normal counter-clockwise rotational effect. The quad-helix appliances when used with brackets and tubes permit alignment of upper incisors after their eruption (Figs. 23B.1; 23B3j–l).

23B.4.2.3 Transverse Expansion

The modified quad-helix appliance is removed from the teeth and adjusted at 6-week intervals (Fig. 23B.2a, b). A force of about 200 g on each side is

optimal. The arch increases in width approximately 3 mm per month, irrespective of cleft type [18]. The cuspid segments often need more expansion than the molar area. Because there is a great tendency for relapse, overexpansion is necessary.

With transverse expansion of the maxillary arch, a downward clockwise rotation of the mandible occurs without any forward movement of the maxilla [18]. By comparing the use of fixed quad-helix appliances with removable expansion plates on noncleft patients, Hermanson et al. [42] found that a fixed quad-helix appliance was more effective with fewer visits, less costly, and required shorter treatment time. A removable plate would not readily resist the forward-downward traction to the cuspid area from a facial mask. As a rule, transverse expansion is completed before protraction is started.

23B.4.2.4 Protraction

The quad-helix appliance is formed to make passive contact with the incisors, by bends or by a soldered-on extension (Fig. 23B.2 a,b). In cases where no transverse expansion is needed, or if the quad-helix spring is inconvenient after the expansion period, a simple palatal arch is soldered to four bands on second deciduous molars and cuspids (Fig. 23B.2 e,f). The intraoral appliance is used as anchorage for the facial mask (Scheu: Great Lake Reversed Pull Face Crib, 2500.1 small) (Fig. 23B.2 c,d). No other mask fixation is needed other than the two intraoral elastics [Unitek: Latex ex-oral 1/4" LGT; Unitek/3M: "Fran" 8 oz. 1/4" (404-736)] from hooks in the cuspid areas. The force used for facial protraction is about 350 g on each side, totaling 700 g. The facial mask is used mainly at night for 10–12 h. Sleeping disturbances have not been reported. Patient cooperation is excellent in almost all cases. Within a few days, the children manage to put on the mask themselves, and complaints about soreness are very rare. It is important that the elastics be attached to the anterior segment, and inclined downward and forward about 15° to the occlusal plane. Several facial masks of various designs are now available. If the use of protraction forces is delayed until the permanent incisors are fully erupted, elastic forces from hooks placed on the arch wire between the lateral incisor and cuspid area may be utilized. In most cases, the incisors should be advanced bodily to obtain surface bone deposition at subnasal (A-point) by use of edgewise arch wires with labial root torque (Fig. 23B.3 l).

23B.4.2.5 Fixed Retention

The results obtained by early orthopedics must be stabilized with a passive fixed palatal arch wire. The retainer remains until the anchor deciduous teeth are lost (Fig. 23B.2 e). The palatal arch wire (or quad-helix appliance) may be combined with multibracketed appliances to align the permanent incisors (Fig. 23B.2 f).

Treatment Timing of Interceptive Orthopedics

Proper treatment timing during the late deciduous dentition or early mixed dentition periods is of utmost importance. Delaire [22, 23] observed that maximum skeletal maxillary changes occurred when protraction therapy was instituted before 8 years of age. Tindlund [16, 17] found significantly better skeletal response when protraction began at 6 years (mean age 6.3). At this age the annual sutural growth rate is nearly as high as that found at the pubertal period [43], when development of the heavily interdigitated sutural systems has already commenced [44, 45]. Protraction during the deciduous dentition period minimizes unwanted dentoalveolar proclination of maxillary incisors in the permanent dentition [10, 18].

Early habilitation of facial appearance and dental functions, preferably before the start of school, is considered a major goal [8, 13, 14]. The cooperation of the young patients is often more predictable at this age [16, 17]. The objective of having the permanent upper incisors erupt into a positive overjet and overbite relationship warrants that orthopedics should be started even earlier in cases with severe skeletal jaw discrepancies. Postponement of orthopedic treatment increases the likelihood that achieving positive effects on the facial growth pattern will fail to occur.

23B.4.3 Treatment Results of Using a Protraction Facial Mask

23B.4.3.1 Clinical Results

The excellent results achieved by the early use of interceptive orthopedics to correct an anterior and/or posterior crossbite also have a desirable effect on improving dentofacial appearance [9–11, 16–19, 22, 23, 46]. Treatment outcomes can vary because success also depends on inherited craniofacial characteristics and the severity of the primary and secondary midfacial malformations related to clefting. Generally, the treatment response is less favorable in facial types with increased anterior vertical height and a steep mandibular plane angle.

Patient cooperation is of major importance for obtaining a good treatment outcome. Using protraction therapy in 108 CLP patients with anterior crossbite in the deciduous dentition, Tindlund et al. [10] achieved favorable incisor relationships in 98 cases. Significant increase of maxillary skeletal prognathism by protraction was found only in the UCLP group, whereas treatment effects in the BCLP cases were mainly dentoalveolar [11]. The observation of significant differences between the UCLP and BCLP groups in Bergen is most likely associated with the primary surgical procedures utilized, which included a periosteoplasty. A bony fusion of jaw segments in BCLP on one or both sides may impair treatment response as well as facial growth.

After protraction treatment there was no significant difference in the maxillary prognathism attained between the UCLP and BCLP groups [11]. The sagittal position of the upper molars was normalized in both groups. Increase of the upper facial height (n-sp") and clockwise rotation of the occlusal plane were significantly greater in the BCLP group. The upper incisors were still retroclined in both groups, which is considered a beneficial state. A later dentoalveolar proclination will compensate for future mandibular development. On the average, the period of protraction lasted 12 months in the UCLP group and 15 months in the BCLP group.

The skeletal response to maxillary protraction is expected to vary considerably as a consequence of skeletal facial variation, differences in the cleft defects, and in cleft repair [16]. Favorable response in the sagittal skeletal maxillo-mandibular jaw relationship was found in 63% (mean increase of angle ANB was 3.3°), whereas favorable response on skeletal forward movement of the maxilla was found in 44% (mean advancement 2.4 mm). A combined favorable response of both the mandible and the forward movement of the maxilla was found in 35% [16]. In this group the mean increase of the maxillary prognathism was 2.1°, the angle ANB increased 3.7°, the maxilla moved forward 3.1 mm, and the maxillary dentition was advanced 4.3 mm. In cases where the sagittal jaw discrepancy was due to overgrowth of the mandible, the resulting changes accentuated a mandibular downward/posterior rotation, increasing anterior facial height.

Cephalometric predictors for good orthopedic treatment response were retrusion of the upper jaw due to short maxillary length resulting in a Class III skeletal and dental relationship and counter-clockwise inclination of the occlusal plane. This is associated with a retrusion of the upper lip and the nose tip [17]. Favorable increase of a positive ANB-angle is associated with mandibular retrognathism, whereas skeletal forward movement of the maxilla with lesser

changes in the ANB angle was more often seen in cases with normal mandibular prognathism.

23B.4.3.2 Limitations

During protraction the upper permanent incisors should never be proclined beyond the supporting basal bone, and the lower incisors should never be retroclined more than their normal position within the alveolus [10]. If a normalization of the maxillo-mandibular skeletal discrepancy is not achieved along with normal dental axial inclinations of the permanent incisors, further protraction should be avoided and orthognathic surgery considered (see Categories 2A, 2B in Sect. 23B.4.4). On the other hand, protraction during the deciduous dentition is advocated in every case with an anterior crossbite, even in cases with a family history of true mandibular prognathism. The final diagnosis for orthognathic surgical treatment should be delayed until approximately 13 years of age for girls and the late teens for boys.

23B.4.3.3 Stability/Relapse

After protraction the maxilla and mandible appear to maintain their original growth pattern. Although there is no relapse of the corrected upper jaw relationship [15], the maxillomandibular relationship often worsens through normal forward growth of the mandible while the maxillary position relative to the anterior cranial base appears to remain constant. However, long-term results show individual variation of this finding, and in cases with moderate midfacial retrusion early protraction is often sufficient to maintain the improved inter-incisor relationship with growth (Fig. 23B 1) [47].

23B.4.3.4 Soft-Tissue Profile

As already stated, the characteristic concave profile with midfacial retrusion is readily improved with protraction (Fig. 23B.1) [46]. The changes were nearly the same in BCLP and UCLP patients with significant protrusion of the upper lip (mean increase of 3.0° in the Holdaway angle; mean increase of maxillo-mandibular lip positioning (SS-N-SM, angle of 2.5°) [19]. Although there is a close relationship between the soft-tissue profile and the supporting hard-tissue structures [19, 48], the improved soft-tissue profile commonly seen after protraction is more stable than the ANB-angle which is also dependent on mandibular position, size, and growth [15].

23B.4.4 Long-Term Prognosis After Interceptive Orthopedics

By age 6 years the craniodentofacial growth and development may give some indications of the facial appearance at adulthood. The CLP patients with few exceptions can be placed into one of four categories, irrespective of the type of clefting [47]:

Category 0: Normal skeletal facial morphology.

Need of treatment:

- Alignment of upper permanent incisors at age 7–8 years?
- Conventional orthodontic treatment at age 11–13 years.

Prognosis: Very good.

Category 1: Normal skeletal facial morphology, except posterior crossbite(s).

Need of treatment:

- Interceptive orthopedics: Transverse expansion of the upper jaw at age 6–7 years.
- Alignment of upper permanent incisors at age 7–8 years?
- Conventional orthodontic treatment at age 11–13 years.

Prognosis: Very good.

Category 2A: Moderate skeletal facial discrepancies.

Need of treatment:

- Interceptive orthopedics: Transverse expansion and protraction of the upper jaw at age 6–7 years.
- Alignment of upper permanent incisors at age 7–8 years?
- Conventional orthodontic treatment at age 11–13 years.

Prognosis: Good/fair for a permanent result.

Category 2B: Severe skeletal facial discrepancies, however, cannot be differentially diagnosed from Category 2A until age 12–15 years

Need of treatment:

- Interceptive orthopedics: Transverse expansion and protraction of the upper jaw at age 6–7 years.
- Alignment of upper permanent incisors at age 7–8 years?
- Conventional orthodontic treatment at age 11–13 years.
- Combined orthodontic/orthognathic-surgical correction at adult age.

Prognosis: After orthognathic surgery: Good regarding upper arch form, tooth position, and soft tissue profile. Poor permanent result until after orthognathic surgery with stable retention of the arch form.

Patients in Category 2B may be candidates for orthognathic surgery at an adult age. This is not a contraindication for early maxillary protraction provided this does not lead to dentoalveolar compensations that undermine the possibility for optimal surgical intervention at a later age. On the contrary, early orthopedics may create a better physical environment for successful orthognathic surgery. Thus, less maxillary advancement at the time of surgery may improve the postsurgical stability. Besides, the child has greatly benefited from an improved dentofacial appearance during the important formative years.

23B.5 Conclusions

The specific aims of early interceptive orthopedics/orthodontics are:

1. To achieve maximum facial aesthetics, with an adequate incisor overjet–overbite relationship and “a nice dental smile” with symmetry of the dental and facial midlines.
2. The elimination of anterior and posterior crossbite and the recovery of space for the erupting incisors. This is considered “lege artis” (standard operating procedure) in children without clefts, and, obviously, the same considerations are valid for a child with a cleft.
3. Early orthopedic-orthodontic correction generates an optimal skeletal base to accommodate erupting upper permanent incisors and improve dental function.

Protraction produced significant changes: (1) a more anterior position of the upper jaw and (2) a more posterior position of the chin point due to mandibular downward-backward rotation. Significant increase of skeletal maxillary prognathism was found only in the UCLP patients, while in BCLP cases the treatment effect was mainly dentoalveolar.

The initial growth pattern reappears after protraction, with the upper jaw’s position relative to the anterior cranial base remaining stable, while the mandible’s position changes as it grows forward and downward. Soft-tissue profile changes are lasting.

Fixed appliances are indispensable for controlled orthopedic/orthodontic mechanics to obtain all treatment objectives and for the permanent retention of the corrected arch form. Bonded palatal retainers are frequently required.

A diagnosis-related checklist is the method of choice for individualizing orthodontic treatment. Orthopedic/orthodontic intervention should be based on the same principles that are valid for noncleft patients.

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23C LeFort I Osteotomy

S.A WOLFE, SAMUEL BERKOWITZ

23C.1 Surgical Maxillary Advancement LeFort I Osteotomy

Not long ago, maxillary advancement seemed a formidable procedure to many surgeons. Cleft patients with Class III malocclusion often were treated by the more familiar method of mandibular setback, even though the problem, by clinical and cephalometric examination, could be shown to be in the maxilla.

Today, the LeFort I osteotomy is a standard adjunct to the treatment of patients with cleft lip and palate. No matter how gentle or atraumatic the original surgery on the lip and palate, there will probably always be cleft patients who require the LeFort I procedure. It should be as much a part of the armamentarium of cleft palate teams as closure of the lip or palate or a pharyngeal flap.

Mandibular growth should be largely completed before a maxillary advancement is performed; for girls this age is around 14 to 15, and for boys perhaps a year or two older. Most orthodontists advise that several lateral cephalometric films, taken 6 months apart, should show no further growth before the operation is scheduled.

As for timing, it is better to perform the lip and nasal surgery during separate sessions. If the alveolus is intact and there are no buccal crossbites, expansion of the maxilla is not required, and the LeFort I osteotomy is a relatively simple procedure. The nonintubated nostril is packed with cocaine/epinephrine-impregnated gauze, as for rhinoplasty, and the upper labial sulcus is infiltrated with a 1:2,000,000 epinephrine/hyaluronidase solution. The incision is made above the reflection of the sulcus, sparing the frenulum. The mucosal incision does not extend beyond the first molar. Subperiosteal dissection of the anterior maxilla is carried out to the infraorbital rims, visualizing the infraorbital nerves, and then taken posteriorly beneath the mucoperiosteal tunnel to the pterygomaxillary space. If the dissection is strictly

subperiosteal, there is no bothersome exposure of the buccal fat. The piriform aperture is dissected, sometimes removing a portion of the nasal spine, and the nasal mucoperiosteum is dissected back to the hard palate–soft palate junction. The septum can either be separated bluntly from the vomer or a guarded osteotome can be used. The osteotomy is performed largely with the reciprocating saw, starting laterally in the thick bone beneath the buttress of the zygoma and proceeding medially through thinner bone. The osteotomy through the piriform aperture and medial wall of the antrum is done with the saw blade pointed laterally.

Sectioning of the palatine bone, the sole attachment of the maxillary tuberosity to the pterygoid plate of the sphenoid, follows. The lateral osteotomy can be taken a bit farther back by a few taps on a straight osteotome, and the medial antral wall can be further sectioned with a guarded nasal osteotome.

At this point, the only remaining attachment of the lower maxillary segment is the posterior wall of the antrum, and firm, downward finger pressure on the maxilla is usually enough to produce a down-fracture. If not, the forceps can be inserted underneath the nasal mucosa and the maxilla completely mobilized with a downward and side-to-side motion. It can be further mobilized with a blunt elevator used as a lever.

The maxilla is then placed in the desired occlusal relation with the mandible, and both jaws are placed in the desired relationship with the rest of the face. An autogenous iliac or cranial bone graft is used when the face is to be lengthened, when the degree of maxillary advancement is more than 5 mm, or when the patient has a cleft. If the maxilla is shortened, the resected bone is placed over the osteotomy lines.

Sometimes the alveolus is intact, but the maxilla needs to be expanded, as may occur in a cleft patient who has a buccal crossbite and an alveolar cleft. This procedure is easily performed from above the hard



Fig. 23C.1 a-f. Instrumentation for the LeFort I osteotomy. **a** Reciprocating saw with irrigation (Aesculap). **b** Guarded septal and nasal osteotomes. **c** Kawamoto osteotome. **d** Rowe for-

ceps with rubber guard on the palatal arm. **e** Nestor (blunt, heavy, periosteal elevator modified by Jack Nestor Engineering, Inc., Miami, Florida). **f** Expansion forceps

palate, and the palatal mucosa is kept intact if possible. The sectioning is performed with the reciprocating saw, and an elevator is inserted to gently pry the two segments apart. Expansion forceps can be used if required. If the palatal mucosa absolutely prevents expansion, it is divided, creating an alveolar and anterior palatal cleft.

If there is an alveolar cleft to begin with, the two maxillary segments are handled independently and brought into proper occlusion with the mandible. The palatal cleft-nasal floor defect is bone-grafted, and if necessary a transportation flap is developed from the buccal sulcus (Burian) to close the palatal defect. In rare instances, a tongue flap is required. The nasal lining, which will have been carefully dissected at the beginning, is closed before the palatal bone graft is inserted.

The procedure has now been refined to the stage that is the same regardless of whether the alveolus was initially intact. Miniplates are placed between the upper and lower portions of the maxilla for rigid fixation. If bone grafts are required, they are placed either between or over the bone cuts.

If the desired maxillary advancement measures more than 6 mm, bone grafts can be wedged into the pterygomaxillary gap. This step is facilitated by using a traction wire placed through the thick bone beneath the nasal spine. The wire is used to pull the maxilla to the opposite side, which opens the gap and allows impaction of the bone graft. Circumzygomatic wires are almost never used, because they pull the maxilla back, they are too long (long wires can “stretch” more than short wires), and they do not prevent the anterior maxilla from rocking downward.

Wolfe [1] uses an iliac or cranial bone graft on all cleft patients, as these patients are likely to have a maxillary relapse. Generally, the bone can also be used as an onlay to fill out a deficient maxilla. If the advancement is less than 5 mm, bone is placed only over the anterior osteotomies and in the alveolar and palatal cleft, if present.

The use of anything other than a fresh autogenous bone graft is unsafe. It takes about 15 min to harvest the needed amount of iliac or cranial bone. In the former case, the patient will be comfortable as far as the hip is concerned within 1 to 2 weeks. By this time, the autogenous graft will have consolidated. With cadaver or demineralized bone or with hydroxyapatite, consolidation may require months, or may *never* occur.

Like the sagittal splitting procedure for the mandible, the LeFort I osteotomy, once mastered, can provide a solution to a number of maxillary problems. After the horizontal osteotomy, down-fracture, and mobilization, the maxilla can be:

1. Advanced directly with or without a bone graft (in the noncleft class III patient).
2. Advanced, or advanced and expanded transversely, with a bone graft (in the cleft patient).
3. Moved superiorly after resection of a measured amount of maxilla above the horizontal osteotomy (in cases of “long face,” resulting from vertical maxillary excess).
4. Moved inferiorly with a bone graft (in cases of “short face,” or vertical maxillary deficiency).
5. Sectioned into multiple segments with teeth (Wassmund or Schuchardt procedure, done from above).
6. Moved directly backward, although this is difficult to do. (The resection should be of the maxillary tuberosity after extraction of the third molars rather than of the pterygoid plate.) The same result can generally be achieved by an associated segmental osteotomy performed more anteriorly.

With the maxilla in the down-fractured position, multiple osteotomies can be performed from above, which, coupled with or without dental extractions, permit the dental correction of complex malarrangements of the maxilla in one stage. The circulation of blood to the anterior segment comes entirely through the palatal mucoperiosteum, and one must be certain that there are no protrusive edges from the occlusal splint to impinge on the anterior palate. Any number of transverse sagittal osteotomies can be performed, depending on the requirements of the individual case.

Attempts to treat an anterior open bite by mandibular ramus osteotomies are often unsuccessful due to relapse caused by the predominance of the masticatory muscles. Anterior segmental osteotomies of the mandible are appropriate when there is dental crowd-

ing and a downward angulation of the mandibular occlusal plane.

The Schuchardt procedure can be used to shorten posterior maxillary height, but it is rarely used in the USA because it requires either an interdental osteotomy or a tooth extraction (Fig. 23C.2).

If the orthodontist can level the maxillary occlusal plane, even by accentuating the open bite, the simplest and most stable solution is the LeFort I osteotomy. If the position of the maxillary central incisors relative to the lower vermilion border of the upper lip is satisfactory beforehand, this relationship is preserved. If desired, the maxillary incisors can be raised or lowered relative to the upper lip.

After the maxilla has been completely mobilized, intermaxillary fixation is established and the maxillo-mandibular complex seated with firm upward and posterior pressure to set the condyles. Appropriate resection of the posterior and, if necessary, the anterior maxilla is performed until the desired anterior maxillary height is obtained. Stabilization of the maxillary osteotomy is then performed with miniplates, and the intermaxillary fixation, if utilized, is temporarily discontinued to evaluate the occlusal relationship with the patient; head in a fixed position. This examination will reveal whether the condyles were inadvertently pulled out of the glenoid fossae. A Class II relationship indicates that the maxilla must be posteriorly repositioned, either by resecting a portion of pterygoid plates (which is difficult) or by extracting the maxillary third molars and resecting a portion of the maxillary tuberosity (which is easier) (Figs. 23C.2, 23C.3).

23C.2 Stability of Maxillary Advancement

A disappointing yet frequent sequel to orthognathic surgery to advance the maxilla is its partial or complete return to the original state (relapse). The maxillary advancement occurs within a limiting soft tissue envelope (the skin and muscles). Mandibular advancement surgery, especially when it involves the mandibular ligaments, has a great tendency to relapse. The degree of relapse is often judged by measuring occlusal or skeletal landmark changes.

Hochban et al. [2] in a review of the literature, reported that the use of miniplates (in rigid fixations) is superior to wire fixation in overcoming the tendency to relapse. Currently, most reports favor the use of miniplates [4–9]. Proffit and Phillips [10] found a skeletal relapse at 32% after midface advancement using wire fixation compared with 25% after miniplate fixation.

Some investigators believe that the amount of relapse is directly related to the amount of advancement

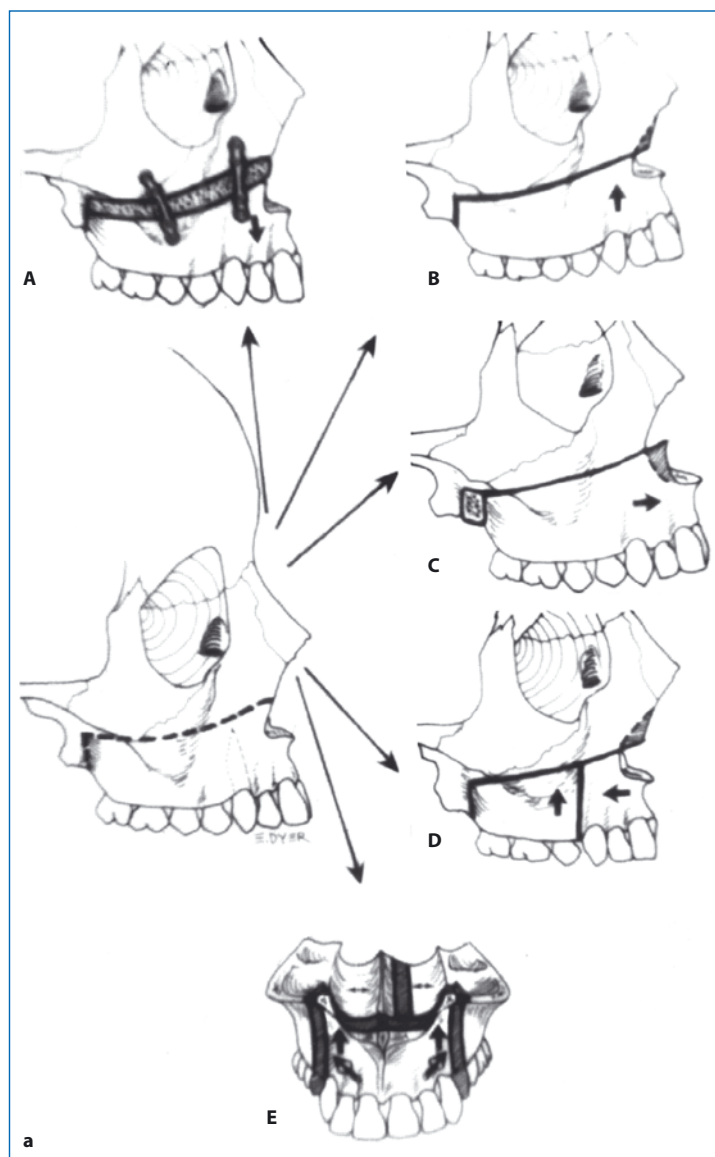


Fig. 23C.2. a LeFort I surgical procedures: Various directions the maxilla can be moved. (A) Inferiorly: requires bone grafts to maintain the new position. (B) Superiorly: No grafts required. (C) Forward: Requires a posteriorly supporting bone graft. (D) The posterior segment is moved superiorly while the anterior segment is moved posteriorly. When the premaxilla is retruded it usually needs to be surgically widened through the midpalatal suture between the central incisors to maintain good cuspid interdigitation. (E) The premaxilla moved superiorly

[1, 5, 11], whereas others think there is no correlation between displacement of the maxilla and relapse [9, 10, 12]. Proffit and Phillips also believe that it is important to achieve excellent occlusion following the operation to reduce the tendency to relapse. Epker [13] suggests that interpositioning of bone grafts increases stability by enhancing bony consolidation.

It is generally accepted that the tendency toward relapse starts immediately after surgery and continues for up to about 6 months after the operation. After about 1 year, the correction can be considered stable [5, 13–15]. Hochban et al. [2], in an excellent review of the subject of postoperative maxillary relapse, reported cephalometric analyses of 31 patients preopera-

tively, postoperatively, and 1 year later. Fourteen patients had clefts of the lip and palate; the others were noncleft patients with maxillary deficiency. All had maxillary advancement by LeFort I osteotomy and miniplate fixation. Hochban et al. [2] found that the amount of relapse was between 20% and 25% in the cleft group and about 10% in the noncleft group. The degree of relapse was related to the amount of advancement, thus confirming the earlier work by Rosen [9] and Houston et al. [5]. The authors recommended surgical overtreatment and a good overbite–overjet relationship after orthodontic treatment.

Berkowitz sometime uses very light Class III elastics for 6 months to improve bony consolidation when

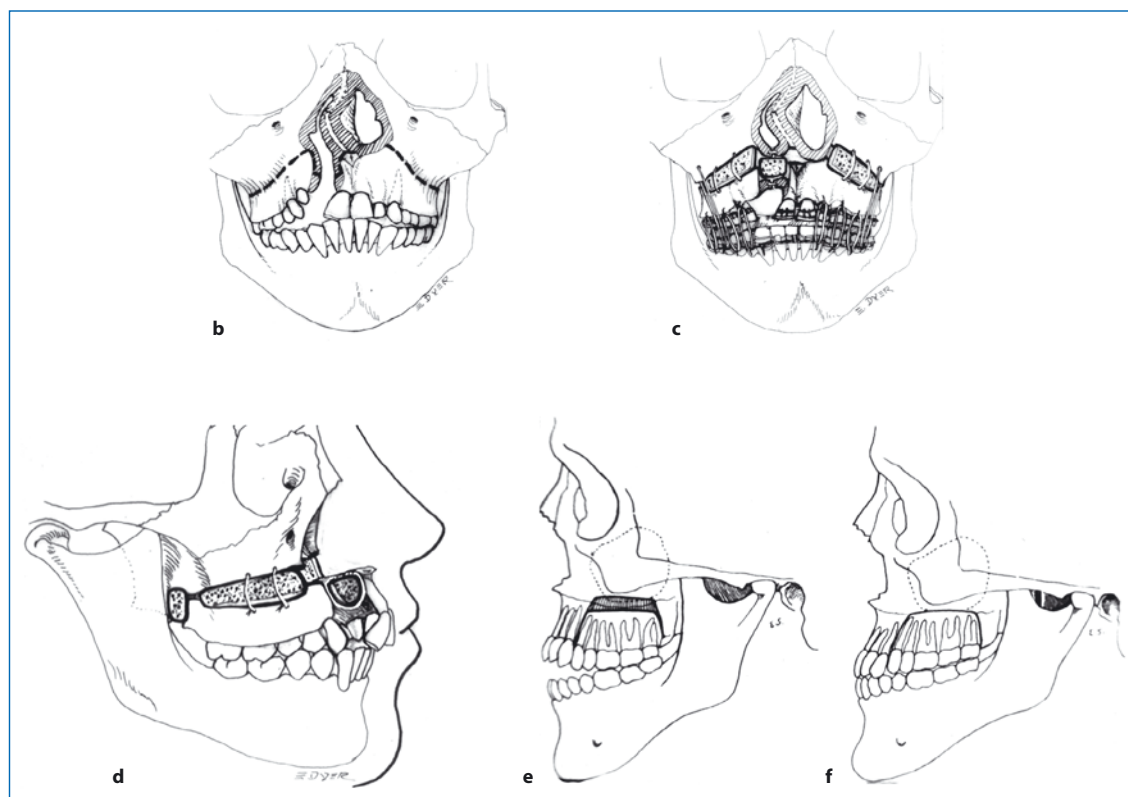


Fig. 23C.3. (continued) **b** Initial incisions for LeFort I surgery with a secondary alveolar bone grafting to be performed simultaneously. **c** The maxilla is moved inferiorly with bone grafts placed at the surgical cite to support the lengthened maxilla. Alveolar bone graft placed from the nasal aperture to the alveolar crest. Prior to the use of metal plates (rigid fixation) steel sutures were used to stabilized the separated segments. An acrylic surgical wafer is used to position the bony segments according to prior mock surgery performed on plaster casts. In-

termaxillary fixation of the maxilla to the mandible using intermaxillary rubber bands for 4–6 weeks is recommended in cases with severe palatal scarring in conjunction with the use of rigid fixation. **d** Lateral view shows a bone block placed between the perpendicular plates of the sphenoid and the maxillary tuberosity with a bone graft to the premaxillary-maxillary junction. **e** Buccal segments are superiorly positioned to permit mandibular auto-rotation and reduction of the anterior open bite

he notices a maxillary relapse occurring. He believes that the muscular drape to the midface changes very slowly in adapting to skeletal changes, and therefore, some overtreatment is necessary in all instances.

Posnick and Ewing [16] studied the outcomes in 30 adults and adolescents judged skeletally mature, who had unilateral cleft lip and palate and underwent LeFort I advancement. This group was investigated to determine the amount and timing of relapse, the correlation between advancement and relapse, the effect of performing multiple jaw procedures, the effect of different types of bone grafts, the effect of pharyngoplasty in place at the time of osteotomy, and the effectiveness of various methods of internal fixation.

Tracings of preoperative and serial postoperative lateral cephalograms were digitized to calculate horizontal and vertical maxillary changes. No significant

differences in outcomes were seen between patients who had maxillary surgery alone and those who had operations on both upper and lower jaws, nor did the outcomes vary significantly with the type of autogenous bone graft used or the segmentalization of the LeFort I osteotomy. Average “effective” advancement was greater both immediately and 2 years after surgery in patients who did not have a pharyngoplasty in place before the operation.

Advancement also was more stable both immediately and 2 years after surgery in the patients with miniplate fixation than in patients with direct-wire fixation. Mean downward (vertical) displacement was 2.6 mm with a relapse of 1.4 mm after 2 years. The degrees of relapse and of advancement or displacement did not correlate significantly.



Fig. 23C.4 a-h. Case JS (AV-64) UCLP showing LeFort I advancement to correct midfacial retrusion. Treatment: Increase midfacial height, and widen the palatal arch. **a-g** Pre- and post-surgical facial and intraoral photographs showing changes in

the profile and occlusion. Chin augmentation is usually contraindicated with midfacial advancement since it may lead to a concave profile after some maxillary relapse. **h** Type of surgery performed

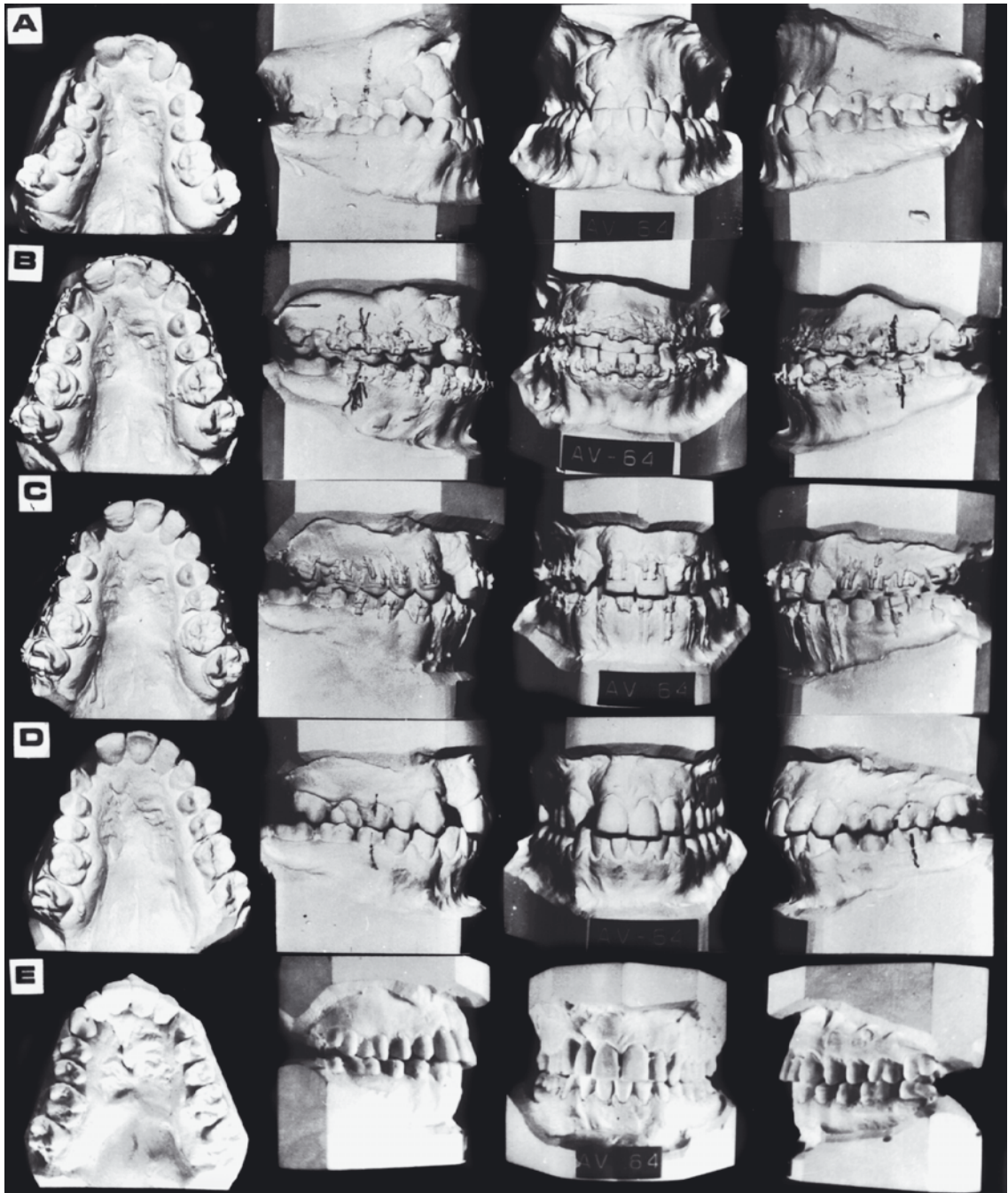


Fig. 23C.5. Case JS (AV-64). Serial dental casts. This case shows severe palatal collapse and scarring leading to buccal and anterior crossbite. Pre- and postsurgical orthodontics plus maxillary surgery reduced the anterior crossbite. The maxillary arch

was orthodontically expanded to open the upper right lateral incisor space and to avoid additional surgery with more palatal scarring

There is another obvious risk factor, the tonicity of the orbicularis oris muscle ring, which needs to be considered. Unfortunately, there are no pressure measurements that can be utilized to improve the success-to-failure ratio.

23C.3 Total Maxillary Advancement and Its Possible Effect on Speech

Jabaley and Edgerton [17] Dez Prez and Kiehn [18], and Bralley and Schoney [19] have reported that speech of cleft and noncleft patients is unaffected after total maxillary advancement. Witzel and Munro [20] say that is not always true. Epker and Wolford [21] noted that the speech of patients with clefts who exhibited no VPI presurgically generally remained unchanged after maxillary advancement. However, those patients who have borderline closure or minimal velopharyngeal incompetence before surgery do exhibit speech changes following total maxillary advancement. Schwarz and Gruner [20–22] showed that patients with slight hypernasality and/or nasal emission before surgery became more hypernasal after maxillary advancement. They concluded that the degree of deterioration was directly related to the extent of maxillary advancement and observed that deterioration could also occur in some noncleft patients. Schendel et al. [23] believe the differences between the two groups are theoretically a reflection of the inherent deficit in palatal musculature and associated soft tissue in the cleft patient and/or cicatrization associated with surgical repair of the palatal clefts. Many cleft patients have hypoplastic velar muscles and associated soft tissues. All of these factors are reflected in the shorter soft palate in the cleft patient. They speculate that the increase in pharyngeal depth creates a significant functional demand which often cannot be met by cleft patients due to less soft palate length increases following maxillary advancement. Schendel et al. [23] believe that the soft palate lengthens about one-half of the amount the maxilla is advanced. They also computed a “need ratio” (pharyngeal depth divided by soft palate length) in which a value of .68 to .84 is consistent with proper velopharyngeal function. A need ratio greater than 1.0 indicates possible postsurgical velopharyngeal incompetence.

23C.4 Technique (Fig. 23C.6)

Unless work is to be done on the nose, a nasal intubation is used. If the premaxilla is absent, an oral tube can be used and simply brought through the central empty space. Schendel and Delaire [23] de-

scribed a technique whereby they brought an oral tube behind the maxillary tuberosity; however, it seems that this method is difficult unless a major advancement is to be performed. It also precludes putting a bone graft in the pterygomaxillary gap. It is better to perform the lip and nasal work surgery at a separate sitting.

If the alveolus is intact, and expansion of the maxilla is not required, the LeFort I osteotomy is an easy procedure. The nonintubated nostril is packed with cocaine/epinephrine-impregnated gauze, as for a rhinoplasty, and the upper labial sulcus is infiltrated with a 1:200,000 epinephrine/hyaluronidase solution. The incision is made above the reflection of the sulcus, sparing the frenulum. The mucosal incision does not extend beyond the first molar. Subperiosteal dissection of the anterior maxilla is carried out to the infraorbital rims, visualizing the infraorbital nerves, and then taken posteriorly beneath the mucoperiosteal tunnel to the pterygomaxillary space using a Cushing elevator. If the dissection is strictly subperiosteal, there is no bothersome exposure of the buccal fat. The piriform aperture is dissected, sometimes removing a portion of the nasal spine, and the nasal mucoperiosteum is dissected back to the hard palate-soft palate junction. The septum either can be separated bluntly from the vomer or a guarded osteotome can be used. The osteotomy is performed largely with the reciprocating saw, starting laterally in the thick bone beneath the buttress of the zygoma and proceeding medially through thinner bone (Fig. 23C.6a,b). The osteotomy through the piriform aperture and medial wall of the antrum is done with the saw blade pointed laterally (Fig. 23C.6 b, c). Section of the palatine bone, the sole attachment of the maxillary tuberosity to the pterygoid plate of the sphenoid, is done with either a curved Dautrey osteotome or the somewhat larger Kawamoto osteotome (Fig. 23C.6d). The lateral osteotomy can be taken a bit further back by a few taps on a straight osteotome, and the medial antral wall can be further sectioned with a guarded nasal osteotome. At this point, the only remaining attachment of the lower maxillary segment is the posterior wall of the antrum, and firm, downward finger pressure on the maxilla is usually enough to produce a down-fracture (Fig. 23C.6e). If not, the Rowe forceps can be inserted underneath the nasal mucosa and the maxilla completely mobilized with a downward and side-to-side motion. It can be further mobilized with a blunt elevator used as a lever (Fig. 23C.6f,g).

The maxilla is then placed in the desired occlusal relation with the mandible, and both jaws are placed in the desired relation with the rest of the face. An autogenous iliac or cranial bone graft is used when the face is to be lengthened, when the degree of advancement is more than 5 to 6 mm, or when the patient has



Fig. 23C.6. Le Fort I osteotomy

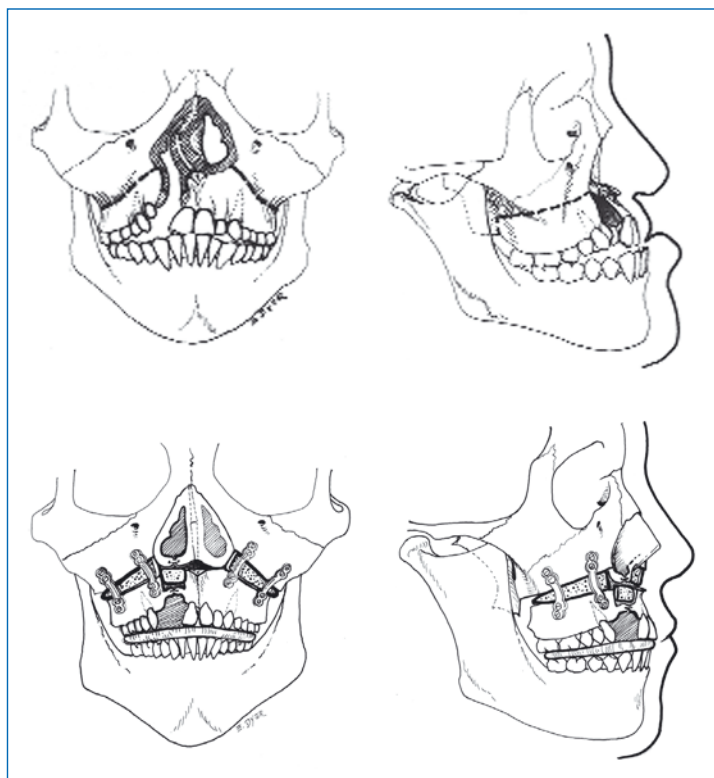


Fig. 23C.7. LeFort I osteotomy in a patient with a unilateral cleft. Autogenous bone grafts are always used, not only to help consolidation and prevent relapse but to improve appearance. I use an iliac or cranial bone graft on all cleft patients, as these patients are likely to have a relapse. Generally, the bone can also be used as an onlay to fill out a deficient maxilla. If the advancement is less than 5–6 mm, bone is placed only over the anterior osteotomies and in the alveolar and palatal cleft, if present. The use of anything other than a fresh autogenous bone graft is unwise. It takes about 15 min to harvest the needed amount of iliac or cranial bone, and the patient will be comfortable as far as the hip is concerned within 1 to 2 weeks. By this time, autogenous grafts will have consolidated. Consolidation with cadaver or demineralized bone or with hydroxylapatite may require 6 months, if indeed it occurs at all. Are we doing the patient a favor here by “sparing” him or her an iliac bone graft? Probably not

a cleft. If the maxilla is shortened, the resected bone is placed over the osteotomy lines.

Sometimes the alveolus is intact, but the maxilla may need to be expanded as in a cleft patient who has had a bone graft of the alveolar cleft. This procedure is easily performed from above, and the palatal mucosa is kept intact if possible. The section is performed with the reciprocating saw, and an elevator is inserted to gently pry the two segments apart. The expansion forceps can be used if required. If the palatal mucosa absolutely prevents expansion, it is divided, creating an alveolar and anterior palatal cleft (Fig. 23C.6).

If there is an alveolar cleft to begin with, the two maxillary segments are handled independently and brought into proper occlusion with the mandible. The palatal cleft–nasal floor defect is bone-grafted, and if necessary a transportation flap is developed from the

buccal sulcus (Burian) to close the palatal defect. In rare instances a tongue flap is required. The nasal lining, which was carefully dissected at the beginning, is closed before the palatal bone graft is inserted (Fig. 23C.7).

The procedure has now reached a stage that is the same regardless of whether the alveolus was initially intact.

Osteosynthesis wires are passed between the upper and lower portions of the maxilla, and suspensory wires are taken through the thick bone beneath the zygomatic buttress laterally.

If bone grafts are required, they are placed either between or over the osteosynthesis anteriorly. If the advancement is more than 5 to 6 mm, bone grafts can be wedged into the pterygomaxillary gap. This step is facilitated by using a traction wire placed through the

Fig. 23C.8. Case JR-AT-94. Midfacial advancement to correct an anterior crossbite and improve facial aesthetics and occlusion. Surgical History: This patient was originally treated in Honduras. Lip closure at 2 months, and palatal closure at 8 months of age. No attempt was made to close the palatal fistulae. Oral Facial Evaluation: Midfacial hypoplasia with an anterior and right buccal crossbite. Treatment Plan: Advance and expand both dental arches and close the palatal fistulae when

surgically advancing the maxilla. Results: Although the patient had a cleft of the lip and palate with twinned deciduous lateral incisors in the line of the cleft, one of the permanent lateral incisors was well developed, the other malformed. The malformed impacted tooth was extracted, a secondary alveolar cranial bone graft, and fistula closing procedure were all performed at the same time (13–11). A LeFort I maxillary advancement brought the teeth into Class I relationship

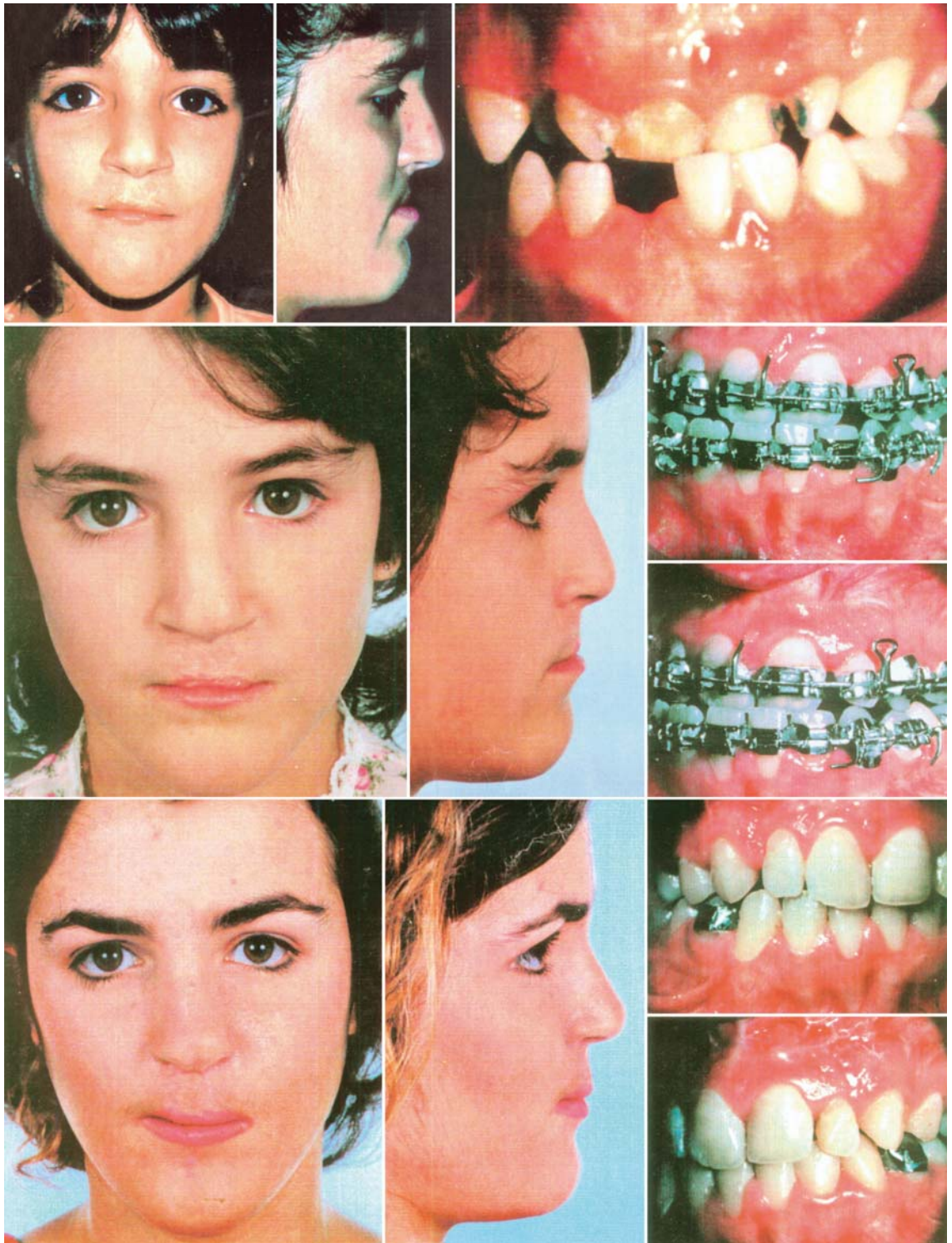


Fig. 23C.8.

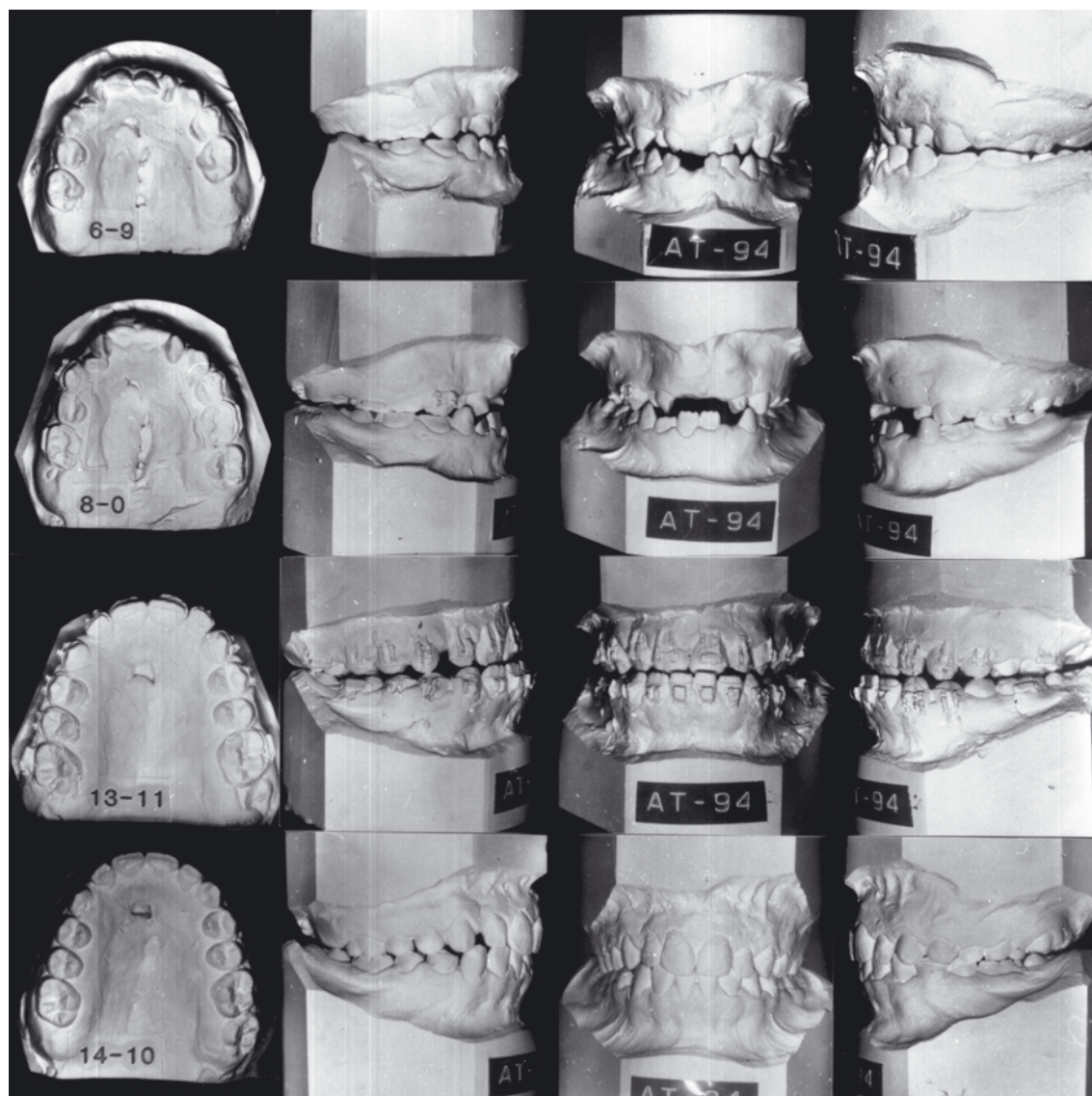


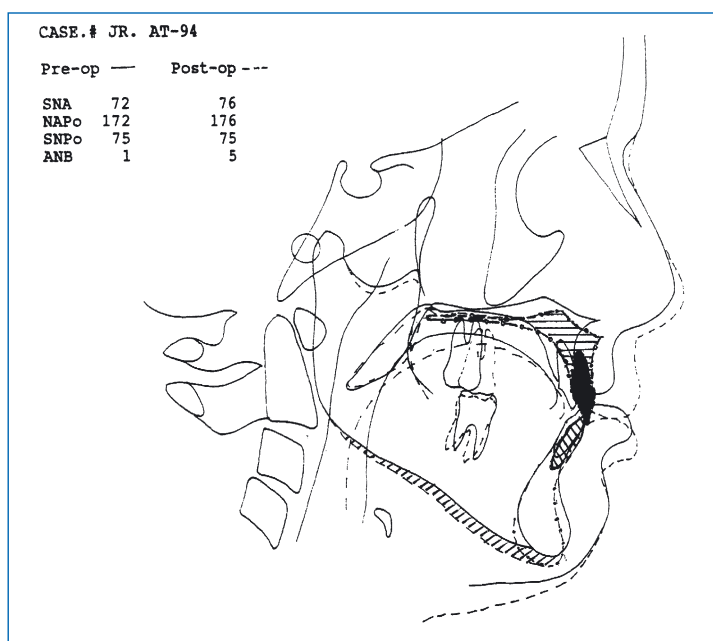
Fig. 23C.9. Case JR (AT-94). Surgical maxillary advancement (LeFort I). Pre- and postmaxillary advancement

thick bone beneath the nasal spine. The wire is used to pull the maxilla to the opposite side, which opens the gap and allows impaction of the bone graft. The suspensory wires from the buttress are brought through the inferior mucosal flap and wired to the upper arch bar. If further suspensory wires are needed, they can be brought down from the infraorbital rim. Circumzygomatic wires are almost never used: They pull the maxilla back, they are too long (long wires can “stretch” more than short wires), and they do not prevent the anterior maxilla from rocking downward.

Like the sagittal splitting procedure for the mandible, the LeFort I osteotomy, once mastered, can provide a solution to a number of maxillary problems. After the horizontal osteotomy, down-fracture, and mobilization, the maxilla can be:

1. Advanced directly with or without a bone graft (nonleft class III patient).
2. Advanced, or advanced and expanded transversely, with a bone graft (cleft patient).
3. Moved superiorly after resection of a measured amount of maxilla above the horizontal osteotomy (long face: vertical maxillary excess). Differential

Fig. 23C.10. Case JR. Lateral cephalometric tracings before and after LeFort I maxillary advancement and inferior positioning. The mandible autorotated posteriorly reducing the mandibular prominence. Maxillary advancement moved the upper lip forward while the lower lip remained practically in the same position



upward movement can also be used to close an anterior open bite.

- Moved inferiorly, with a bone graft (short face: vertical maxillary deficiency).
- Sectioned into multiple segments with dental extractions: Wassmund or Schuchardt procedure from above.
- Moved directly backward. It is difficult to do (the resection should be of the maxillary tuberosity after extraction of the third molars rather than of the pterygoid plate). The same result can generally be achieved by an associated segmental osteotomy performed more anteriorly.

23C.5 Multiple Maxillary Osteotomies

With the maxilla in the down-fractured position, multiple osteotomies can be performed from above, which, coupled with dental extractions, permit correction of complex malarrangements of the maxilla in one stage. The circulation to the anterior segment comes entirely through the palatal mucoperiosteum, and one must be certain that there are no protrusive edges from the occlusal splint to impinge on the anterior palate. Any number of transverse or sagittal osteotomies can be performed depending on the requirements of the individual case.

Class III malocclusion following isolated cleft palate repair. The patient had a large palatal cleft that was closed at 18 months of age with a von Langenbeck

repair. Because of severe velopharyngeal insufficiency, a bipedicle island flap incorporating both greater palatine arteries was positioned at age 7, along with a palatal pushback. Following this procedure speech has been normal, but the patient showed subsequent restriction of maxillary growth, both transversely and in the posteroanterior direction. After preliminary orthodontic expansion of the maxilla, a LeFort I osteotomy was done at age 16. The maxilla was advanced without difficulty and stabilized in the usual fashion with iliac bone grafts. The mucosa beneath the horizontal incision at the end of the procedure was pale blue, which is not unusual in a cleft patient. However, over the next few days the upper portion of the mucosal flap proceeded to slough in a way that suggested lack of blood supply more than infection. Healing eventually occurred, with partial obliteration of the upper labial sulcus and partial exposure of the roots of the central incisors, which required periodontal treatment. Despite vigorous and sustained postoperative orthodontic treatment, the occlusion relapsed to a Class III relation.

Figure 23C.10 shows a patient who has had a unilateral palatal island flap can safely undergo a LeFort I osteotomy, but as this patient demonstrates, it is not a safe procedure in one who has had diversion of both greater palatine arteries. Either the mandible should be moved back or a LeFort II osteotomy performed in which most of the anterior vestibular mucoperiosteum is preserved. Figure 23C.10a,b shows a raw area of the anterior palate following placement of a bipe-

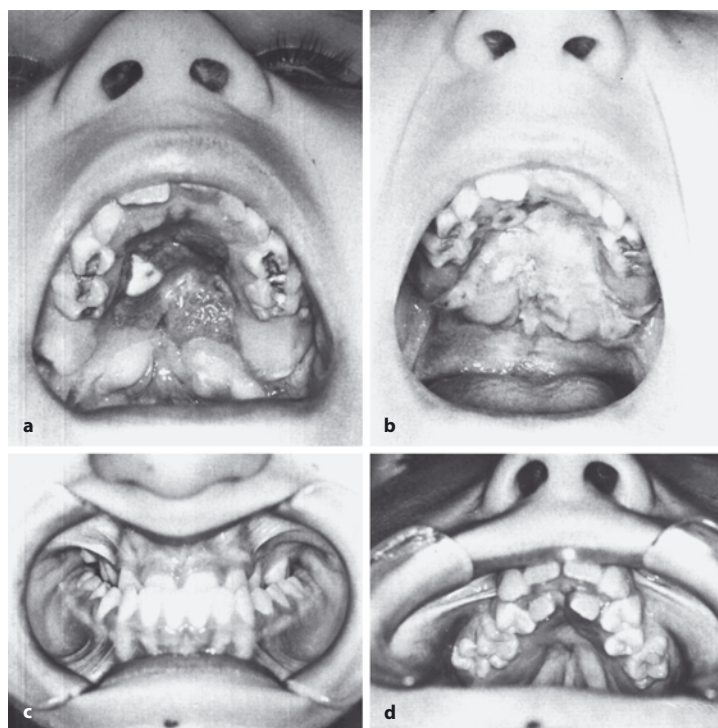


Fig. 23C.11 a–j. Class III malocclusion following isolated cleft palate repair. The patient had a large palatal cleft that was closed at 18 months of age with a von Langenbeck repair. Because of severe velopharyngeal insufficiency, a bipedicle island flap incorporating both greater palatine arteries was positioned at age 7, along with a palatal pushback. Following this procedure speech has been normal, but the patient showed subsequent restriction of maxillary growth, both transversely and in the posteroanterior direction. After preliminary orthodontic expansion of the maxilla, a LeFort I osteotomy was done at age 16. The maxilla was advanced without difficulty and stabilized in the usual fashion with iliac bone grafts. The mucosa beneath the horizontal incision at the end of the procedure was pale blue, which is not unusual in a cleft patient. However, over the next few days the upper portion of the mucosal flap proceeded to slough in a way that suggested lack of blood supply more than infection. Healing eventually occurred, with partial obliteration of the upper labial sulcus and partial exposure of the roots of the central incisors, which required periodontal treatment. Despite vigorous and sustained postoperative orthodontic treatment, the occlusion relapsed to a Class III relationship. Fig. 23C.10 shows a patient who has had a unilateral palatal island flap can safely undergo a LeFort I osteotomy, but as this patient demonstrates, it is not a safe procedure in one who has had diversion of both greater palatine arteries. Either the mandible should be moved back or a LeFort II osteotomy performed in which most of the anterior vestibular mucoperiosteum is preserved. **a, b** A raw area of the anterior palate following placement of a bipedicle island flap showing rapid healing by secondary epithelialization. **c, d** Subsequent maxillary deformity 5 years later. Note the transpalatal scar band

dicle island flap showing rapid healing by secondary epithelialization. Figure 23C.10c,d shows subsequent maxillary deformity 5 years later. Note the transpalatal scar band.

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Fig. 23C.11. (continued) **e–j** Before and after LeFort I osteotomy. Note the appearance of the gingiva above the maxillary incisors. **k** There is partial exposure of the roots of the central incisors. **l, m** Degree of relapse after 6 months, with partial obliteration of the upper labial sulcus

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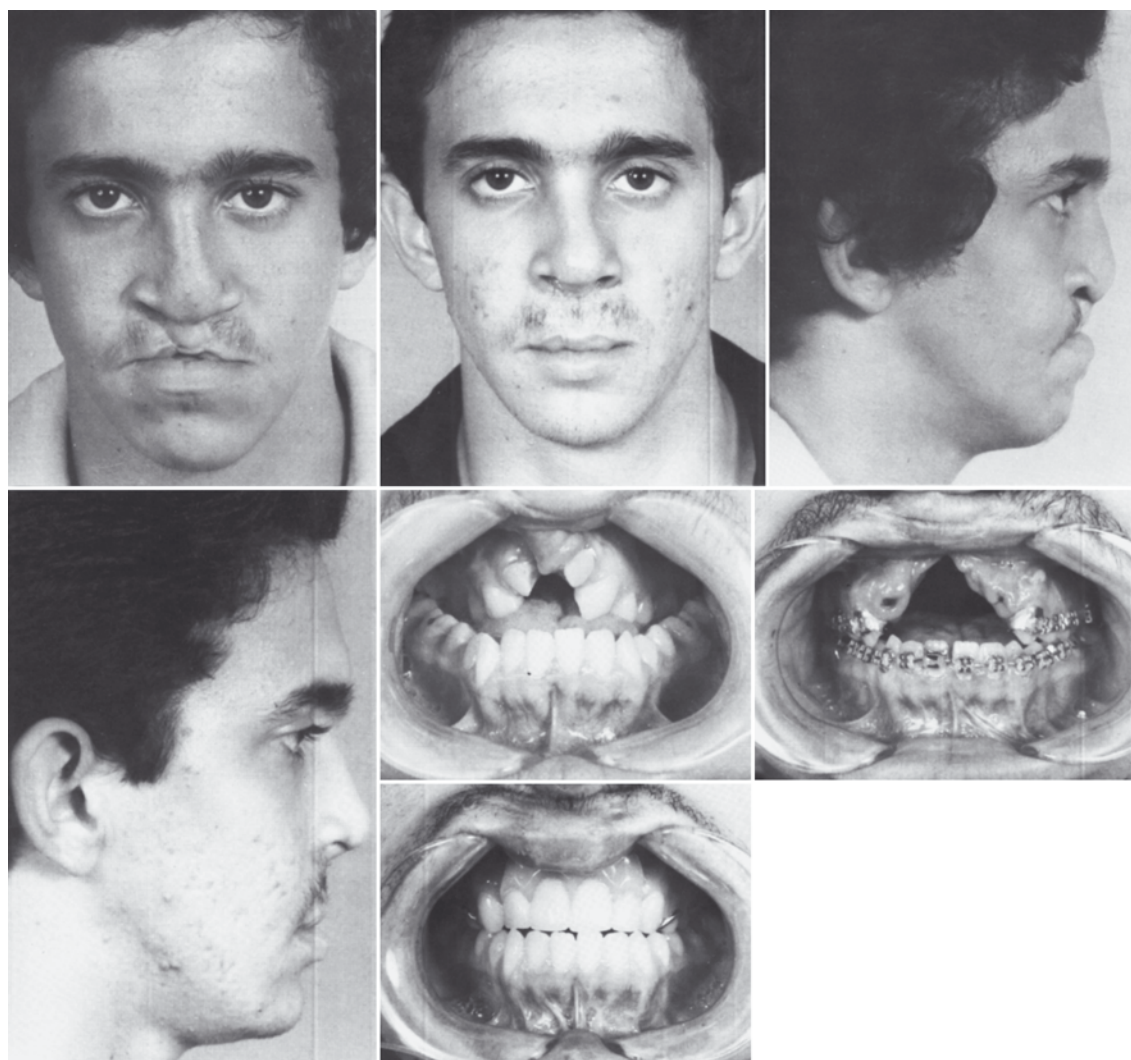


Fig. 23C.12. Bilateral cleft with absent premaxilla. Original treatment had been administered in Cuba, and the premaxilla had been excised at the time of the lip closure. Severe maxillary collapse ensued. A LeFort I osteotomy and maxillary expansion

was performed, with bone grafting of the palatal cleft. A prosthesis replaced the missing teeth. Following this, Ralph Millard proceeded with a cleft lip rhinoplasty and Abbe flap

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23D Rigid External Distraction: Its Application in Cleft Maxillary Deformities

JOHN W. POLLEY, ALVARO A. FIGUEROA

Patients with severe maxillary deficiency secondary to orofacial clefting present multiple challenging problems for the reconstructive team. These patients exhibit multidimensional maxillary hypoplasia and skeletal clefting with absence of maxillary and alveolar bone, as well as scarring, residual fistulas, and dental anomalies. Traditional surgical/orthodontic approaches to treat these patients, while sometimes successful in obtaining stable occlusal relationships, often fall short of expectations with respect to facial balance and aesthetics. The application of maxillary distraction osteogenesis in the treatment protocol of patients with severe maxillofacial anomalies offers a powerful alternative for the reconstructive team.

In this chapter we present the use of maxillary distraction osteogenesis for the treatment of patients with severe maxillary deficiency secondary to orofacial clefts. The technique of rigid external distraction, based upon the old concept of cranial fixation, has enabled rigid control over the distraction process allowing for predictable and highly successful outcomes.

23D.1 Materials and Methods

All patients seen at our institution between April 1, 1995, and December 1, 1996, with severe maxillary hypoplasia (negative overjet of 8 mm or more) were considered candidates for treatment with maxillary distraction osteogenesis. Criteria for patient selection included the following: unilateral or bilateral cleft lip and palate, severe maxillary hypoplasia (vertical, horizontal, and transverse planes) with class III malocclusion, patients requiring horizontal maxillary advancement in excess of 8 mm, normal mandibular morphology and position, patients with full primary dentition or older, patients with severe palatal and pharyngeal scarring, and patients with airway obstruction and sleep apnea.

Table 23D.1. Number of patients by diagnosis and gender

Diagnosis	n	Male	Female
UCL/P	10	6	4
BCL/P	6	5	1
Facial Cleft	2	1	1
TOTAL	18	12	6

UCL/P, unilateral cleft lip and palate; BCL/P, bilateral cleft lip and palate.

Eighteen consecutive patients were selected based on the above criteria and were treated with maxillary distraction osteogenesis. There were 10 unilateral cleft lip and palate patients, 6 bilateral cleft lip and palate patients, and 2 patients with bilateral cleft lip and palate and severe congenital facial clefting (Table 23D.1). The patients' ages at the time of surgery ranged from 5.2 years to 25.2 years.

All patients underwent a thorough history and clinical examination as well as a complete dental and orthodontic examination. Preoperative and postoperative photographic and cephalometric records were obtained as reported previously. Time was spent with the patient and the patient's family, explaining the distraction process in detail utilizing photographs, video imaging, as well as discussions with other patients and their families who have undergone the procedure. The patient and parents were thoroughly familiarized with the distraction apparatus and its mechanics prior to the procedure.

For this group of patients a custom-made intraoral orthodontic splint, which acts as the link between the maxillary skeleton and distraction apparatus, was inserted in each patient (Fig. 23D.1). This intraoral splint was constructed with rigid 0.045 or 0.050 stainless steel orthodontic wire. The splint was cemented to the first permanent molars or second primary molars and was further secured at the time of surgery with

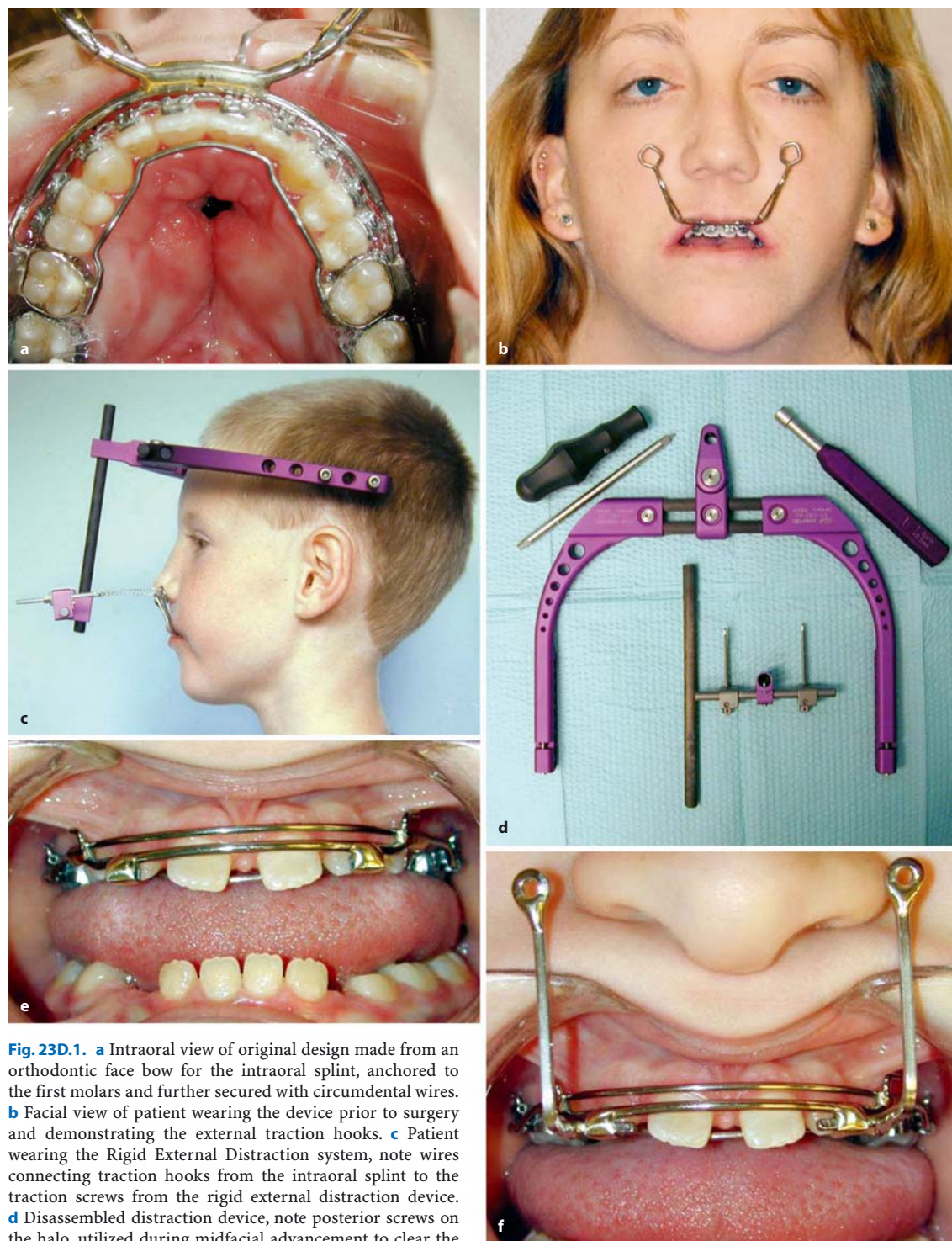


Fig. 23D.1. **a** Intraoral view of original design made from an orthodontic face bow for the intraoral splint, anchored to the first molars and further secured with circumdental wires. **b** Facial view of patient wearing the device prior to surgery and demonstrating the external traction hooks. **c** Patient wearing the Rigid External Distraction system, note wires connecting traction hooks from the intraoral splint to the traction screws from the rigid external distraction device. **d** Disassembled distraction device, note posterior screws on the halo, utilized during midfacial advancement to clear the anterior part of the halo from forehead. **e** Intraoral view of newly designed intraoral splint with removable external distraction hooks. Designed in this fashion to avoid the presence of the hooks at surgery. **f** Intraoral splint with removable distraction hooks in place

circumferential stainless steel surgical wire. For older children and adults, the splint was inserted in the clinic prior to the surgical procedure. For young children, the splint was placed in the operating room after anesthetic induction and prior to the osteotomy. External traction hooks were bent under and in front of the upper lip with the end of the hook ending at the level of the palatal plane. The splint also included intraoral hooks that were utilized for the required retention phase at the completion of active maxillary distraction. Certain orthodontic tooth movements as well as orthodontic expansion of the upper arch can be performed at the time of maxillary distraction by incorporation of an expansion screw and segmentalizing the intraoral splint into two units. In order to improve rigidity the splint can be manufactured by using an orthodontic cervical face bow. The external bow is bent to create the external traction hooks. Currently the intraoral part of the splint utilized at our center is made of heavy .045 or .050 stainless steel orthodontic wire. In the anterior region two square tubes are soldered just medial to the oral commissures (Fig. 23D.1e). These tubes are used to house removable traction hooks made of heavy rectangular wire (Fig. 23D.1f). This design offers the advantage of not having the external traction hooks present at the time of surgery facilitating anesthesia management and intraoral surgical manipulation.

All 18 patients in this series underwent a Le Fort I osteotomy with the height of the transverse osteotomy varying depending on the dental age and aesthetic requirements of each patient. In patients with transitional dentition, a high transverse maxillary osteotomy was performed, just below the level of the inferior orbital rim. The osteotomy circumvents the infra-orbital foramen to prevent injury to the infraorbital neurovascular bundle and avoids all permanent tooth buds. Mobilization of the maxillary segments is achieved, including pterygomaxillary and septal disjunction. No intraoperative repositioning of the maxillary segments is performed, and no autogenous or alloplastic bone grafting or internal skeletal fixation hardware is utilized. For the patients in this series who underwent rigid external distraction, the halo portion of the distraction device was placed immediately after closure of the intraoral incision. In the first four consecutive patients with severe cleft maxillary hypoplasia that we elected to treat with maxillary distraction, we performed the distraction with elastic traction as described below. After four consecutive treatment failures in these patients, we abandoned elastic distraction for rigid external distraction.

Fourteen patients underwent rigid external maxillary distraction (Fig. 23D.1, Table 23D.2) [1]. These patients had the rigid device placed at the time of surgery and had their distraction performed through mechanical activation of the distraction device. Four

Table 23D.2. Treatment groups by diagnosis

Distraction	UCL/P	BCL/P	BCL/P +FC	Total
Rigid external	9	3	2	14
Face mask	3	1	–	4

UCL/P, unilateral cleft lip and palate; BCL/P, bilateral cleft lip and palate; FC, facial cleft.

patients in this series underwent face mask elastic distraction osteogenesis (Table 23D.2). An orthodontic face mask was utilized with elastics placed on the intraoral hooks. Up to 2 lb of elastic force was utilized for each patient through a combination of 8-oz elastics. Patients were instructed to wear the face mask 24 h a day for a period of 3 months. All patients underwent a latency period of 4–5 days following the osteotomy and then began distraction. For the rigid external distraction group, distraction was performed at the rate of 1 mm/day. In this group, the distraction device was kept in place 3–4 weeks following completion of the distraction process for rigid retention. An additional 4–6 weeks of face mask elastic retention at night time only was utilized in this group. For the rigid external distraction group following the period of rigid retention, the external distraction device was removed in an office setting. For the very young patients in this series, the distraction device was removed with mild sedation.

23D.2 Cephalometric Evaluation

The preoperative and postretention lateral cephalometric radiographs were utilized for analysis. The radiographs were traced on 0.003-in acetate paper, and 12 anatomic landmarks were recorded (Fig. 23D.2). All tracings were done by a single experienced investigator (A.A. Figueroa). Availability of serial radiographs in all patients permitted landmark verification. All x-rays were corrected to 0% magnification. Based on the recorded anatomic landmarks, 13 measurements were calculated, 6 angular, and 7 linear (4 horizontal and 3 vertical). For the linear measurements, an x-y coordinate system utilizing the S-N plane as the horizontal was employed. Linear horizontal changes were measured relative to a line perpendicular to the S-N plane passing through sella, and vertical changes were measured perpendicular to the S-N plane. The preoperative and postoperative cephalometric values in the rigid external distraction group were statistically analyzed by means of a paired t-test. The numbers in the face mask distraction group were too small for meaningful statistical analysis for intra-group and intergroup comparisons.

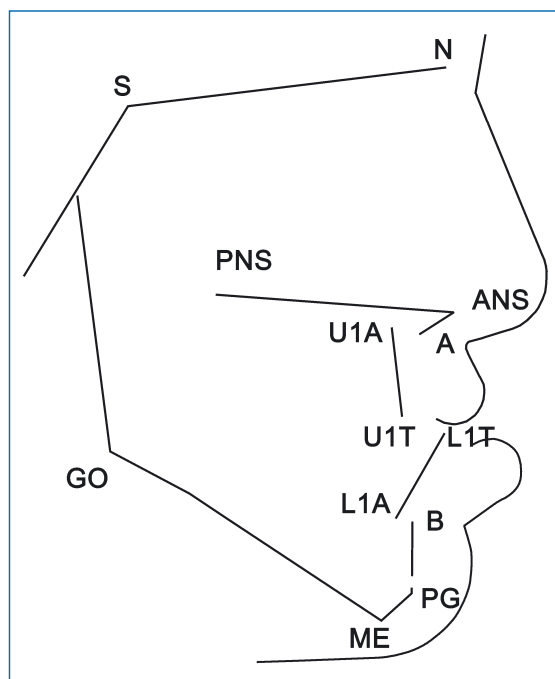


Fig. 23D.2. Anatomic landmarks and reference planes. Anatomic landmarks: sella (S), center of sella turcica; nasion (N), most anterior point of the nasal frontal suture; anterior nasal spine (ANS), most anterior point of the spine; A point (A), most anterior limit of the maxillary alveolar bone at the level of the incisor root apex; posterior nasal spine (PNS), intersection between the nasal floor and the posterior contour of the maxilla; apex of maxillary incisor root (U1A), uppermost point of the incisor root; tip of maxillary incisor crown (U1T), maxillary incisor edge; tip of mandibular incisor (L1T), mandibular incisor edge; apex of mandibular incisor root (L1A), lowermost point of the mandibular incisor root; B point (B), most anterior limit of the mandibular alveolar bone at the level of the incisor root apex; pogonion (PG), most anterior limit of the mandibular symphysis; gonion (GO), the point at the greater convexity of the mandibular gonial region. Reference planes: sella-nasion plane (SN), palatal plane (line through ANS and PNS), maxillary incisor axis (UI) (line passing through U1A and U1T); and mandibular plane (tangent to the lower border of the mandible through ME and GO)

23D.3 Results

Surgery in this series was performed on either an outpatient or a 23-h admission basis by a single experienced surgeon (J.W. Polley). No patient in this series was discharged later than the morning after surgery. Perioperative antibiotics were routinely used. All patients began routine oral hygiene and an unrestricted soft diet 24 h postoperatively. No intermaxillary fixation was utilized.

There was no surgical morbidity in any of the 18 patients in this series. There were no complications of bleeding or infection. No patients required a blood transfusion, and there were no problems of dental injury, avascular necrosis, or gingival injury. In the patients who underwent rigid external distraction, there were no complications with wearing the external device including pain, discomfort, or loosening of the device during the distraction process. The intraoral splint remained intact in all patients through the active and retention phases. None of the families had difficulty following the guidelines for the distraction, which they carried out at home.

The postdistraction cephalometric radiographs for all patients who underwent either rigid external or face mask distraction were obtained 4 months following distraction. The predistraction and postdistraction angular cephalometric measurements for those patients undergoing rigid external and face mask distraction are given in Table 23D.3. The differences in the linear measurements between the predistraction and postdistraction cephalometric radiographs for each group are given in Table 23D.4. In all of the patients who underwent rigid external distraction, the desired treatment goals were obtained. All patients in the face mask distraction group were undercorrected with residual edge to edge anterior dental relations or remained in anterior crossbite. The results in all patients in the face mask distraction group were considered unsuccessful.

23D.3.1 Angular Changes

For patients undergoing rigid external distraction, the average predistraction S-N-A angle was 77.6°, and the postdistraction S-N-A angle was 85.3°, for an average increase in this group of 7.7° (Table 23D.3). The average predistraction A-N-B was -1.2°, and postdistraction was 7.3° with an increase of 8.6°. For all patients who underwent rigid external distraction, the skeletal angle of convexity increased postdistraction by 17.2°. All of these three measurements were statistically significant.

In the elastic distraction group, the average predistraction S-N-A angle was 77.5°, and postdistraction was 80.3° with only an increase of 2.8°. The A-N-B angle was -5.4° predistraction, and postdistraction averaged -1.0° for an increase of 4.4°. The average change in the skeletal angle of convexity postdistraction in the face mask group was 8.8°. The mandibular plane angle changed 2.2° in the rigid external distraction group and 3.3° in the face mask distraction group.

The angular changes in the rigid distraction group were more than double those in the face mask elastic distraction group.

Table 23D.3. Predistraction and postdistraction angular cephalometric measurements for rigid external and face mask distraction groups

Distraction	Measurements (degrees)	Predistraction	Postdistraction (4 months)	Difference	Significance
Rigid external (<i>n</i> = 14)	SNA	77.6 ± 5.6	85.3 ± 5.6	7.7 ± 2.9	**
	SNB	78.8 ± 4.0	77.9 ± 4.1	-0.8 ± 1.8	NS
	ANB	-1.2 ± 3.5	7.3 ± 3.0	8.6 ± 3.6	**
	Convexity (NAPg)	-3.5 ± 7.5	13.7 ± 6.0	17.2 ± 7.3	**
	Mand. Pl./SN angle	39.2 ± 6.7	41.4 ± 5.9	2.2 ± 2.4	*
	U1-P.PL. angle	100.7 ± 15.7	98.8 ± 14.4	-1.2 ± 11.3	NS
Face mask (<i>n</i> = 4)	SNA	77.5 ± 4.3	80.3 ± 5.5	2.8 ± 4.9	
	SNB	82.9 ± 4.3	81.3 ± 3.3	-1.6 ± 3.8	
	ANB	-5.4 ± 2	-1.0 ± 3.4	4.4 ± 2.7	
	Convexity (NAPg)	-12.2 ± 3.5	-3.4 ± 7.4	8.8 ± 5.5	
	Mand. Pl./SN angle	33.2 ± 4.9	36.5 ± 3.6	3.3 ± 4.3	
	U1-P.PL. angle	105.1 ± 13.1	107.3 ± 10.4	2.2 ± 11.8	

p < 0.01.

** *p* < 0.001.

Table 23D.4. Predistraction and postdistraction linear cephalometric measurements for rigid external and face mask distraction groups

Landmark (axis)	Pre-Post-distraction change rigid external (mm)	Pre-Post-distraction change face mask (mm)
ANS-x	7.1 ± 3.9*	2.9 ± 1.4
ANS-y	-0.4 ± 3.0	-1.3 ± 1.8
A point -x	8.3 ± 3.3*	2.8 ± 2.1
A point -y	-1.3 ± 3.4	-1.7 ± 2.3
U1-x	11.6 ± 4.6*	5.2 ± 2.1
U1-Y	-1.8 ± 3.5	-1.8 ± 1.2
Overjet	12.7 ± 3.0*	7.2 ± 2.0

* *p* < 0.001.

8.3 mm. In the elastic distraction group, the A point advancement was only 2.8 mm. The horizontal advancement at the upper incisal edge averaged 11.6 mm for the patients who underwent rigid external distraction (Table 23D.4) and only 5.2 mm for those patients in the face mask elastic distraction group. Patients in the rigid external group had a positive correction of their overjet by 12.7 mm as compared with 7.2 mm in those patients who underwent face mask elastic distraction. All of the changes in the horizontal linear measurements in the rigid external group were highly significant between predistraction and postdistraction measurements at a *p* value of 0.001.

23D.3.3 Dental Changes

The dental changes in both groups between predistraction and postdistraction cephalograms are also given in Tables 23D.3 and 23D.4. In the rigid external distraction group, the change in the angle of the upper incisor edge to the palatal plane averaged -1.2° for all patients. This was not statistically significant. In the face mask distraction group, the angular change in the upper incisor edge to palatal plane measurements was 2.2°. In none of the patients in this series, including both rigid external distraction and face mask groups, were spaces created posterior to the most distal point of anchorage of the intraoral splint.

23D.3.2 Linear Changes

The A-N-S change between predistraction and postdistraction cephalometric radiographs in the rigid external distraction group was 7.1 mm, and in the face mask distraction group it was only 2.9 mm (Table 23D.4).

In the rigid external group, the average horizontal advancement of the A point following distraction was

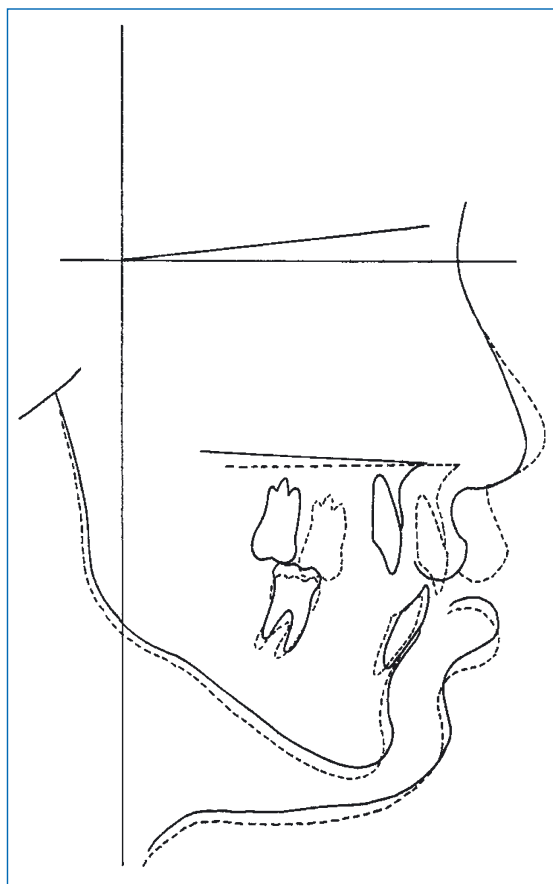


Fig. 23D.3. Predistraction (*solid line*) and postdistraction (*broken line*) average tracings for rigid external distraction group. Note significant maxillary advancement (effective incisor advancement 11.6 mm) with correction of overjet, improvement of skeletal convexity, and minimal changes in mandibular position. Note significant soft tissue changes including the significant degree of lip and nasal tip advancement

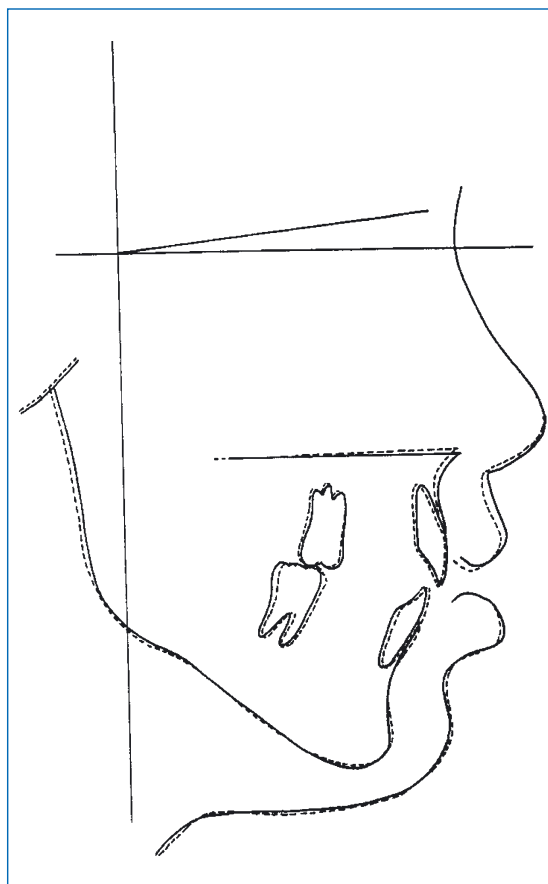


Fig. 23D.4. Postdistraction (*solid line*) and 1 year (*broken line*) after distraction average tracings of the cleft sample treated with the Rigid External Distraction system. Note excellent stability of the advanced maxilla

23D.4 Discussion

Maxillary hypoplasia is a common finding in patients with orofacial clefting. It has been estimated that 25%–50% of all patients born with complete unilateral cleft lip and palate will be potential candidates for maxillary advancement to correct the functional deformities and improve aesthetic facial proportions [2]. In addition, it is recognized that the majority of patients with facial clefts have morphologically normal or slightly smaller than normal mandibles [3, 4]. Patients with severe cleft maxillary deficiency are difficult patients to treat with standard surgical/orthodontic approaches. These patients present with maxillary hypoplasia (vertical, horizontal, and transverse dimensions) and often thin or structurally weak bone.

The maxillary hypoplasia in cleft patients is also compounded by residual palatal and alveolar fistulas, absent and aberrant dentition, and scarring of the palatal and pharyngeal soft tissues.

The physical deformities exhibited by patients with severe maxillary hypoplasia contribute to multiple functional deficiencies as well. These include severe malocclusions, which result in compromised mastication, speech abnormalities, and nasal pharyngeal airway constriction [5]. The severe dish-face or concave facial profiles in these patients result in highly detrimental psychosocial ramifications as well [6]. Current protocols for the treatment of maxillary hypoplasia in cleft patients rely upon a surgical/orthodontic approach, including a Le Fort I maxillary advancement with concomitant fistula closure and maxillary and

Table 23D.5. Review of long-term results of sagittal maxillary relapse in cleft patients following maxillary advancement with fistula closure, bone grafting, and rigid internal fixation

Author	Erbe et al. [7]	Cheung et al. [8]	Posnick et al. [9]	Hochban et al. [10]	Eskenazi and Schendel [11]	Polley and Figueroa [1]
N cleft Pt.	11	46	35	14	12	14
Mean follow-up	59 months	28 months	12 months	12 months	12 months	4 months
Mean max. advancement	4.6	4.5	6.9	8	7.8	11.6
Mean relapse	40%	22%	21%	25%	4%	–

alveolar bone grafting. This surgery includes rigid internal fixation hardware for stabilization of the repositioned maxilla in the postoperative period. The long-term results of cleft patients with maxillary deficiency treated in such fashion have been reported by several authors [7–11]. The mean horizontal maxillary advancement in these reported series has averaged between 5 and 7 mm, and the mean long-term horizontal relapse averages between 20% and 25% (Table 23D.5). Erbe et al. [7] presented a mean follow-up of 59 months for 11 cleft patients who underwent segmental osteotomies with maxillary advancement and simultaneous fistula closure with rigid internal fixation and bone grafting. On average, the greater maxillary segment in these patients was advanced 3.9 mm and the lesser maxillary segment 5.3 mm. At nearly 5 years postoperatively they found that the horizontal relapse of the maxilla was approximately 40%. Cheung et al. [8] reported a consecutive series of 46 cleft patients who underwent maxillary advancement with rigid internal fixation and simultaneous alveolar bone grafting with fistula closure. The mean horizontal maxillary advancement in this series was 4.5 mm. Relapse in the horizontal plane for the unilateral cleft patients in this series, at a mean of 28 months postoperatively, was 22%. Similar horizontal relapse rates following maxillary advancement with rigid internal fixation in cleft patients have been reported by others as well (Table 23D.5) [9–11].

In this series, 14 consecutive patients underwent successful maxillary advancement at the Le Fort I level through maxillary distraction with rigid external distraction. Examples of our clinical experience with rigid external distraction are illustrated in Figs. 23D.5–23D.7. No rigid internal fixation hardware or autogenous bone grafting was utilized at the time of the osteotomy. The patients underwent distraction at the rate of 1 mm per day followed by a 3–4-week rigid retention period. In this group of 14 cleft patients, the mean effective maxillary advancement was 11.7 mm. The time of the postdistraction cephalometric radiographs analyzed in this article averaged 4 months following completion of the distraction process. These data have been presented in this fashion

so that the immediate effects of the distraction process can be measured accurately (Fig. 23D.3). Many of our patients now have a follow-up of more than 12 months, and we have not seen clinical evidence of relapse in any of the patients to date (Fig. 23D.4). In the immature patients, maxillofacial growth will continue, and we are currently following the development of these patients.

In the past, it has been difficult to consistently and successfully treat patients with severe maxillary deficiency with maxillary advancement alone. Patients with large anteroposterior maxillomandibular discrepancies may require mandibular setback surgery in addition to maxillary advancement for correction of their severe horizontal deficiency. With the use of rigid external distraction, we can now gradually and in a very stable fashion reposition a severely hypoplastic maxilla to the exact horizontal and vertical position desired. The patients create their own autogenous bone during this process, eliminating the need for a donor site as well as eliminating the need for rigid internal fixation hardware. The use of rigid external distraction has allowed rigid control over the distraction process and has enabled us to follow our surgical and aesthetic guidelines for the reconstruction of these patients by correcting the entire maxillary skeletal and soft-tissue discrepancy in the region of the hypoplasia only. This expansion of the soft tissue facial mask yields the most pleasing long-term aesthetic facial balance and harmony [12, 13], particularly in cleft patients.

In this initial consecutive series of patients, we utilized two different techniques for maxillary distraction. One group (face mask distraction) underwent maxillary distraction with the use of an orthodontic face mask and elastic traction [14]. With the use of the elastic distraction, we were able to obtain only partial correction of the horizontal maxillary deficiencies in these patients. We considered all of our face mask patients to have failed their surgical treatment. We therefore changed the design of the distraction technique to rigid external distraction, which utilizes a skeletally (cranial) fixed distraction device that allows for rigid, predictable control over the distraction process.

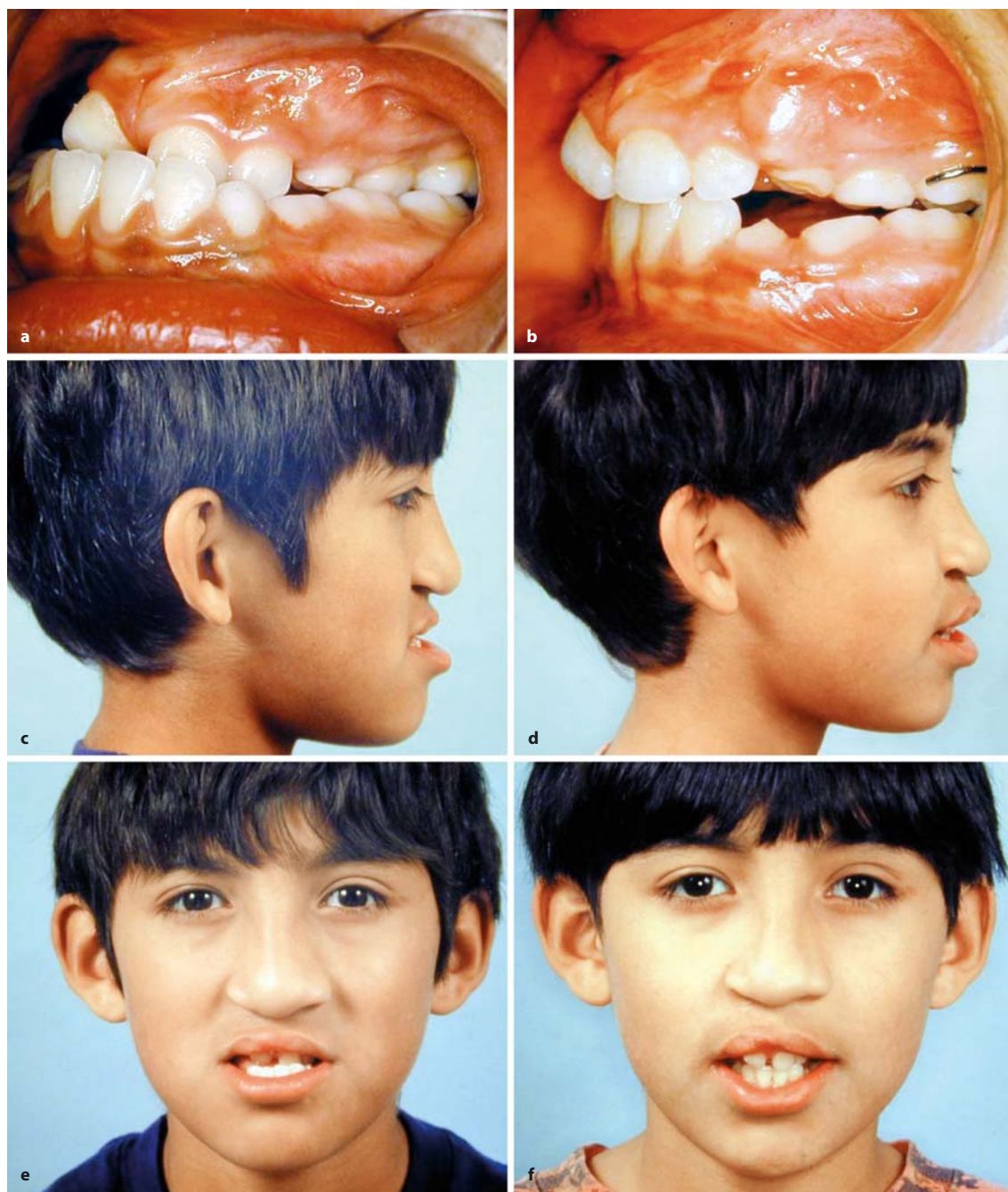


Fig. 23D.5 a–f. Nine-year-old boy with right unilateral cleft lip and palate with severe maxillary hypoplasia and crossbites. Pre-distraction (*left*) and postdistraction (*right*) facial and intraoral

views demonstrate correction of midface deficiency with improved facial proportions and dental relations

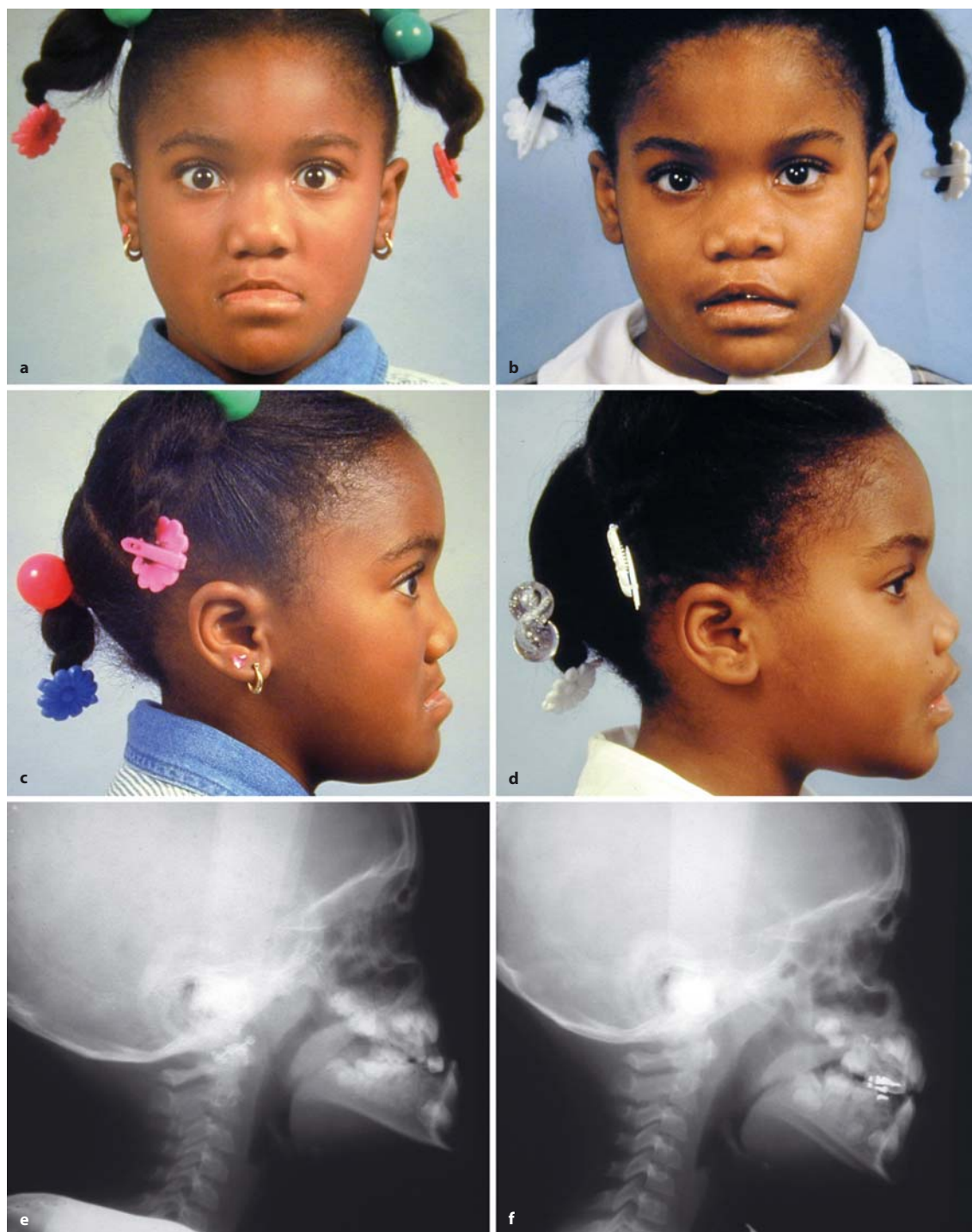


Fig. 23D.6 a-f. Six-year-old girl with left unilateral cleft lip and palate and severe maxillary hypoplasia. Predistracted (*left*) and postdistracted (*right*) facial and cephalometric radiograph views demonstrate correction of facial concavity and crossbite

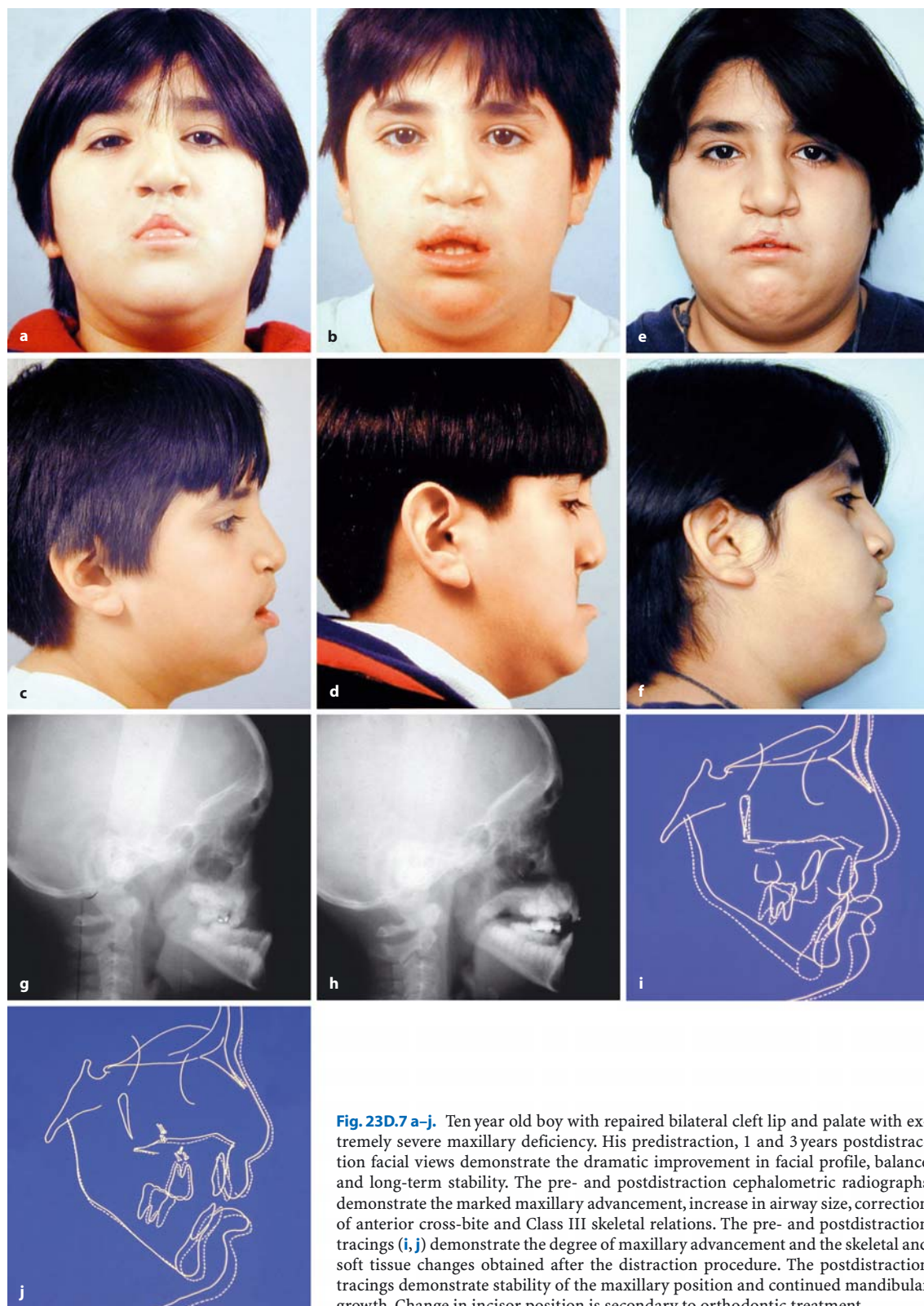


Fig. 23D.7 a–j. Ten year old boy with repaired bilateral cleft lip and palate with extremely severe maxillary deficiency. His predistriction, 1 and 3 years postdistriction facial views demonstrate the dramatic improvement in facial profile, balance and long-term stability. The pre- and postdistriction cephalometric radiographs demonstrate the marked maxillary advancement, increase in airway size, correction of anterior cross-bite and Class III skeletal relations. The pre- and postdistriction tracings (i, j) demonstrate the degree of maxillary advancement and the skeletal and soft tissue changes obtained after the distraction procedure. The postdistriction tracings demonstrate stability of the maxillary position and continued mandibular growth. Change in incisor position is secondary to orthodontic treatment

Table 23D.6. Review of maximum-maxillary sagittal advancement in cleft patients undergoing maxillary distraction with face mask elastic traction

Measurements	Rigid external Chicago	Face mask Chicago	Face mask Rambam, M.C. [15] Haifa	Face Mask C.H.A. Trousseau, [16, 17] Paris
A point advancement (mm)	8.3	2.8	4.5	3
ANB (degrees)	8.6	4.4	4.0	–
Convexity (degrees)	17.2	8.8	5.0	–

This device is readily adjustable, offering the ability to change the vertical and horizontal vector of distraction at any time during the distraction process. The rigid control over the distraction process with rigid external distraction is evidenced by the greater maxillary movements obtained with this device as compared with those patients who underwent face mask elastic distraction. For the patients who underwent rigid external distraction, the mean effective horizontal maxillary movement at the upper incisal edge averaged 11.7 mm. This mean horizontal effective maxillary movement in the face mask elastic traction group was only 5.3 mm. Similarly in the rigid external distraction group, the overjet correction for each patient averaged 12.7 mm versus 7.2 mm for the elastic traction group. The preoperative and postoperative angular cephalometric measurements for each group showed similar results. The average change in the S-N-A angle in the patients who underwent rigid external distraction was 7.7° versus 2.8° in the face mask elastic distraction group. The change in the skeletal angle of convexity in the group who underwent rigid external distraction averaged 17.2° versus 8.8° in the patients who underwent distraction utilizing elastics.

Similar findings in the amount of skeletal maxillary movement with the use of face mask elastic distraction have been reported by others (Table 23D.6) [15–17]. Rachmiel et al. [15] reported that in their series of 12 patients with cleft maxillary deficiency, 30% of the patients were unresponsive to maxillary distraction techniques with face mask elastics. In the 70% of patients in whom they were able to obtain maxillary advancement, the cephalometric findings are similar to our findings in the amount of maxillary skeletal change seen with face mask elastic traction. Rachmiel et al. [15] report the amount of maxillary advancement at the A point in their series to be 4.5 mm, and in our series with face mask elastic traction the amount of movement at the A point was 2.8 mm. A point advancement in our series of patients with rigid external distraction averaged 8.3 mm. The change in the skeletal angle of convexity in the Rachmiel et al. [15] series averaged 5.0° compared with a change of 8.8° in our series of face mask elastic traction and 17.2° in our series of rigid external distraction.

Diner et al. [16] obtained only 3 mm of horizontal maxillary advancement at A point in their series of patients who underwent maxillary distraction osteogenesis with use of a face mask and elastic traction. The experience of Hung et al. [17] was similar with the use of elastic forces for maxillary advancement in cleft patients. In their series, they found the mean horizontal maxillary advancement to be only 5.2 mm. Maxillary distraction at the Le Fort I level with the use of a face mask and elastic traction is unpredictable and unreliable, allowing for horizontal advancement in the 4- to 6-mm range only. In our practice, patients who require only 4–6 mm of maxillary advancement are not routinely considered for treatment with distraction osteogenesis. In these patients, standard orthodontic surgical approaches at the appropriate age are generally performed.

Internal distraction devices have also been reported for use in midface advancement. Most of these reports have focused on advancements of the midface at the Le Fort III level or above [18, 19]. Some, however, have advocated the use of an internal device for maxillary advancement at the Le Fort I level [18]. To date, there is no evidence that this technique affords sufficient distraction at the Le Fort I level. Internal devices may require multiple surgical approaches for their placement, a second operative procedure for their removal, and the necessity for an exit port for the activating arm of the device. The vectors of distraction with the use of internal devices will be limited by the placement of each device as well as its finite mechanics. Perhaps the greatest disadvantage of internal devices is in the design of the transverse maxillary osteotomy. With internal distraction devices, the osteotomy must be designed so that there is enough stable bone above and below the osteotomy line to allow appropriate placement and fixation of the distraction hardware. This is not required with rigid external distraction. The osteotomy design with this device is based upon the aesthetic requirements of each individual patient and not based upon the placement of internal hardware. This allows the transverse maxillary osteotomy to be carried as high as indicated along the pyriform aperture as well as in the malar regions, allowing for maximal correction of the patient's pre-

operative facial concavity. Maxillary advancement with rigid external distraction allows total versatility and flexibility in both the amount and direction of the distraction process. The vectors of distraction can be and are changed at any time during the distraction process. In addition, no additional surgical procedure is required for removal of the rigid external device.

The use of the cranium as an anchorage point for the stabilization of maxillofacial surgery is not a new concept. Cranial stabilization devices for maxillofacial trauma reconstruction as well as for elective maxillofacial surgery have been reported with good success in the past [20, 21]. In addition, our neurosurgical colleagues have used the cranium as a solid fixation point for the stabilization of cervical injuries and reconstructions for decades. The scalp pins (2–3 per side), which stabilize the halo component of the rigid external distraction device, are discomfort free. Not even the youngest patients have any complaints or problems with wearing of the device throughout the distraction process. No special scalp pin care is required, and the use of ointments and creams at the scalp pin interface is discouraged. The patients simply shampoo and wash their hair in the shower with the device in place in a normal fashion. Currently, the device is readily removed without even local anesthesia in the clinic following the phase of rigid retention. In young apprehensive patients the halo is removed with mild sedation. No secondary surgical procedure is required for removal of internal hardware.

One of the great advantages of distraction osteogenesis in the craniomaxillofacial skeleton is that, in theory, there is no limitation to the age at which patients can be treated. Contemporary surgical orthodontic approaches for the treatment of maxillary deficiency in cleft patients are dependent upon the patient having reached skeletal maturity before the reconstructive surgery can be performed. Treating patients in the transitional dentition stage with the use of autogenous bone grafting and rigid internal fixation plates and screws is technically difficult to perform without injury to the developing permanent tooth buds. The technique of rigid external distraction eliminates the negative technical factors associated with traditional orthognathic surgery in patients in transitional dental development. With maxillary rigid external distraction, only an osteotomy is performed. Repositioning skeletal segments, internal fixation hardware, and bone grafting are not required. The only limitations with rigid external distraction include adequate dentition, either primary or secondary, for fixation of the intraoral splint, as well as the ability of the patient to wear the device. Rigid external distraction now allows treatment of these patients from the age of 2 years and up. In special circumstances in patients

with craniosynostosis (Crouzon and Apert syndromes) we have used the technique for monobloc advancement as early as 18 months of age. In these cases, the second or even the first primary maxillary molar have been used to support the intraoral splint. In addition, special plates have been developed to connect the edentulous maxilla to the rigid external distraction device [22].

This article reports our preliminary experience with this technique. Three-year follow-up of this group of patients has demonstrated outstanding stability in maxillary position [23]. However, we caution all young patients undergoing rigid external distraction that they may require a final “finishing” Le Fort I procedure at skeletal maturity for final arch alignment.

The advantages of maxillary rigid external distraction are numerous. This technique allows an excellent modality for correcting the severe midface concavities, at any age, in patients with facial clefting and other hypoplastic anomalies (ectodermal dysplasia, Johansson Blizzard syndrome, etc.). Rigid external distraction follows the important principals of aesthetic maxillofacial surgery treating only the affected jaw and offers the multiple benefits of distraction osteogenesis, including not only orthotopic bone creation with expansion of the facial skeleton but soft-tissue expansion as well. The technical advantages of rigid external distraction are numerous. The surgery is minor as compared with traditional orthognathic surgery. Only the osteotomy is performed, eliminating repositioning of skeletal segments, bone grafting, splints, intermaxillary fixation, and intern fixation hardware. Operative times are significantly decreased. Morbidity with rigid external distraction is very low (in our series zero). Blood transfusions are not required. Rigid external distraction can be performed on an outpatient or 23-h admission basis. Patients begin a soft diet and normal oral hygiene the morning following surgery. The rigid external device is removed in the clinical setting or with mild sedation for the young children. All of the above has resulted in a significantly decreased cost of care for these challenging patients.

In patients with severe cleft maxillary hypoplasia, distraction osteogenesis with rigid external distraction now offers the ability to fully restore facial convexity through a minimal procedure at almost any age. In our practice, this has dramatically changed our treatment philosophies and success for these patients, who in the past have been extremely difficult to treat. The technique is currently applied with extreme success to those patients with severe maxillary nonsyndromal dentofacial deformities, as well as patients with severe syndromic conditions such as Apert, Crouzon and Pfeiffer syndrome [24].

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24 Management of Maxillary Deformities in Growing Cleft Patients

ERIC J.W. LIOU, PHILIP K.T. CHEN

24.1 Introduction

The maxillary deformities in growing cleft patients could be a hypoplastic maxilla, downward and/or laterally displaced premaxilla in bilateral clefts, wide alveolar cleft and fistula, or combinations of the above deformities. It is a challenge for both orthodontist and surgeon to treat a hypoplastic maxilla with wide alveolar cleft in a growing unilateral or bilateral cleft patient, or a downward or laterally displaced premaxilla with wide alveolar cleft and fistula in a growing bilateral cleft patient.

Several new clinical techniques based on the principles of distraction osteogenesis, either nonsurgical orthopedic or surgical approaches, have been developed for managing the maxillary deformities in growing cleft patients. These techniques are:

1. *Effective maxillary orthopedic protraction* for the treatment of hypoplastic maxilla [1] and for minimizing alveolar cleft [2] in unilateral or bilateral cleft patients
2. *Premaxillary orthopedic intrusion* for the correction of a downward displaced premaxilla in bilateral cleft patients [2, 3]
3. *Premaxillary orthopedic medial repositioning* for the correction of a laterally displaced premaxilla in bilateral cleft patients [2]
4. *Interdental distraction osteogenesis* for the approximation of a wide alveolar cleft in unilateral or bilateral cleft patients [4]

24.2 Orthopedic Management of Hypoplastic Maxilla in Growing Unilateral or Bilateral Cleft Patients

The combined use of rapid maxillary expansion and facemask is a contemporary orthopedic management for maxillary protraction in cleft patients [5–7]. It is assumed that rapid maxillary expansion disarticu-

lates the circumaxillary sutures so that facemask protraction of maxilla could be easier [8–10]. Rapid maxillary expansion is sutural expansion osteogenesis of the intermaxillary suture and has been recognized as a form of distraction osteogenesis [11]. Facemask protraction of maxilla is sutural protraction osteogenesis of the circumaxillary sutures. In the absence of a completely intact intermaxillary suture in cleft patients, disarticulation of the circumaxillary sutures through rapid maxillary expansion would play an important role in maxillary protraction.

However, it remains controversial to what width the expansion should reach to disarticulate the circumaxillary sutures. In noncleft patients, some reported that 5 mm of expansion is good enough [12], while some others reported at least 12 to 15 mm [13, 14]. Both for the cleft and noncleft patients, it seems that wider expansion exerts greater tension/stress on the circumaxillary sutures and disarticulates maxilla better than a smaller expansion. However, the expansion should be to displace the maxilla anteriorly and to disarticulate circumaxillary sutures rather than to overexpand the maxilla. To disarticulate circumaxillary sutures without overexpansion, a technique called effective maxillary orthopedic protraction has been developed for orthopedic management of the hypoplastic maxilla in growing cleft patients [1].

24.2.1 Effective Maxillary Orthopedic Protraction

This technique includes three parts:

1. A double-hinged rapid maxillary expander for a greater amount of maxillary anterior displacement
2. A protocol of *Alternate Rapid Maxillary Expansions and Constrictions* (Alt-RAMEC) of maxilla for a better disarticulation of maxilla
3. A pair of intraoral maxillary protraction springs for noncompliant maxillary protraction

24.2.1.1 Double-Hinged Rapid Maxillary Expander

Several types of rapid maxillary expanders have been used for maxillary protraction. They are the *fan-type* [15, 16] or *hyrax-type* built with two acrylic resin halves [8], splints [9], or in a hygienic design [17]. These expanders expand the maxilla in a V-shaped manner [18] with a center of rotation around the posterior nasal spine [19, 20]. The expansion force distributes not only in the maxilla but also into the circumaxillary structures [21, 22]. It is postulated that this would entail bone resorption behind the maxilla and consequently result in posterior displacement of maxilla [17] (Fig. 24.1 a, b). In contrast, it is postulated

as well that this would entail the circumaxillary structures such as pterygoid plates to displace the maxilla forward [23, 24] (Fig. 24.1 c).

These two assumptions explain why some of the clinical studies on hyrax-type expanders reported anterior displacement of maxilla [8, 25, 26], while some others reported no significant displacement [27, 28] or posterior displacement of maxilla [29, 30]. The posterior displacement of maxilla compromises the maxillary protraction in cleft patients.

The double-hinged rapid maxillary expander (US Patent No. 6334771 B1) is developed for a greater amount of anterior displacement of maxilla [1, 2]. Its configuration is similar to a W-appliance and has two hinges of rotation. It consists of a jackscrew in

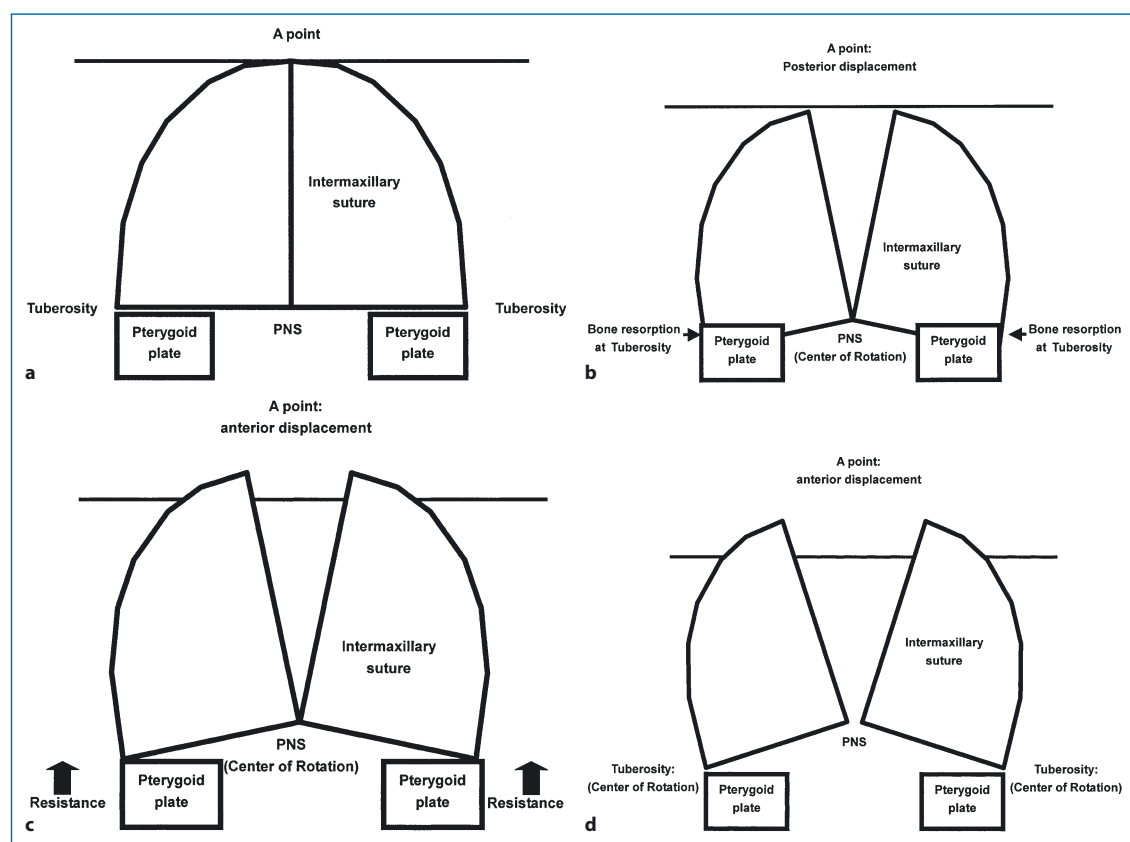


Fig. 24.1 a–d. Schematic illustrations of the postulated maxillary displacement during rapid maxillary expansion. **a** The maxilla before expansion: the *semicircles* represent the right and left maxillae; the *rectangles* represent the pterygoid plates. **b** Posterior displacement of the maxilla after expansion by a hyrax expander: each half of the maxilla rotates outward and backward around the posterior nasal spine (PNS), which entails bone resorption behind the maxillary tuberosities and results in posterior displacement of maxilla. **c** Anterior displacement of the maxilla after expansion by a hyrax expander: each half of

the maxilla rotates outward and backward around the PNS, which entails the circumaxillary structures to displace the maxilla forward and results in anterior displacement of maxilla. **d** Anterior displacement of the maxilla after expansion by a double-hinged expander: each half of the maxilla rotates outward and forward around the maxillary tuberosities, which geometrically results in anterior displacement of maxilla without the possibility of entailing bone resorption behind the maxillary tuberosities

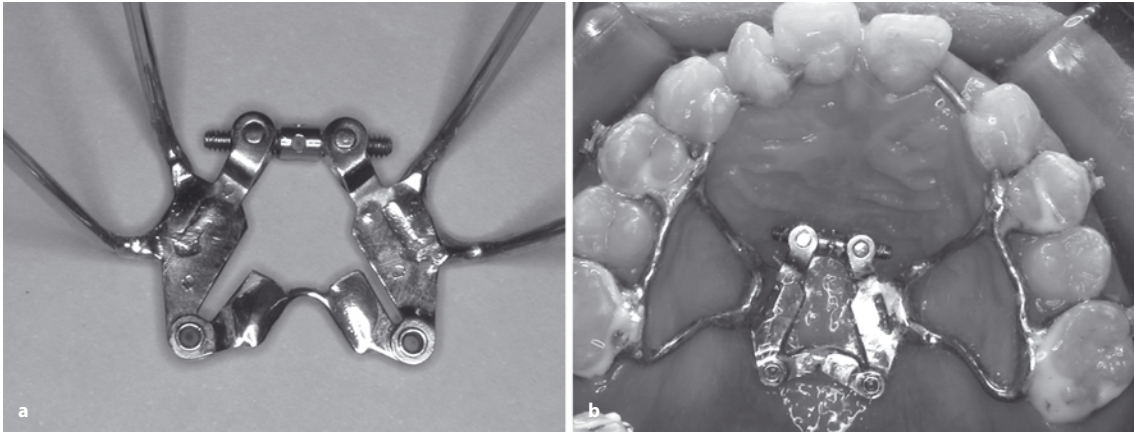


Fig. 24.2 a, b. The configuration of the double-hinged rapid maxillary expander (a), and the assembly of the device in a clinical case (b)

the center, two bolts holding the screw, a body holding the bolts at anterior and two hinges of rotation at posterior (Fig. 24.2a,b). The rationale for its greater amount of anterior displacement of maxilla is it expands and rotates each half of the maxilla laterally and anteriorly through the two hinges of rotation located beside the molars bilaterally. This kind of expansion entails the circumaxillary structures to displace the maxilla anteriorly with less possibility of bone resorption behind the maxillary tuberosity (Fig. 24.1 d).

Maxillary first premolars and molars are banded and maxillary impression is taken for the fabrication of the double-hinged expander. The expander is oriented perpendicular to the central incisors and is soldered to the molar and premolar bands. Two anterior extension arms (0.051-in stainless steel wires) extend bilaterally from the premolar bands toward central incisors (Fig. 24.2b). The premolar and molar bands and the anterior extension arms are sandblasted before cementation. After cementation of the expander, the anterior extension arms are bonded to the anterior teeth with composite resin. One day after cementation, the double-hinged expander is activated according to the protocol of Alt-RAMEC.

24.2.1.2 Alternate Rapid Maxillary Expansions and Constrictions (Alt-RAMEC) of Maxilla

The Alt-RAMEC is a protocol of repetitive weekly alternate rapid maxillary expansions and constrictions [1, 2] (Table 24.1). It lasts for 7–9 weeks until the maxilla is loosened. The weekly (every 7 days) sequence is 7-mm of expansion, 7-mm of constriction, 7-mm of

Table 24.1. Clinical protocol for alternate rapid maxillary expansions and constrictions

Alternate weekly sequence	Weekly amount of expansion/constriction	Daily amount of activation
Expansion	7 mm	1 mm
Constriction	7 mm	1 mm
Expansion	7 mm	1 mm
Constriction	7 mm	1 mm
Expansion	7 mm	1 mm
Constriction	7 mm	1 mm
Expansion	7 mm	1 mm
Constriction	7 mm	1 mm
Expansion	7 mm	1 mm

expansion, 7-mm of constriction, 7-mm of expansion, 7-mm of constriction, 7-mm of expansion, 7-mm of constriction, and 7-mm of expansion etc. Each day the maxilla is expanded or constricted 1 mm/day (four turns in one activation). This protocol allows better disarticulation of circumaxillary sutures without overexpansion of maxilla. Its rationale is similar to simple tooth extraction in which we repeatedly rock the tooth buccally and lingually until the tooth is disarticulated out of the alveolar socket.

Patients are seen once a month. The maxilla is examined clinically for its loosening by holding the patient's head with one hand and rocking the expander with maxilla up and down with another hand. The maxilla is ready for protraction only once loosening of the maxilla has been observed clinically.

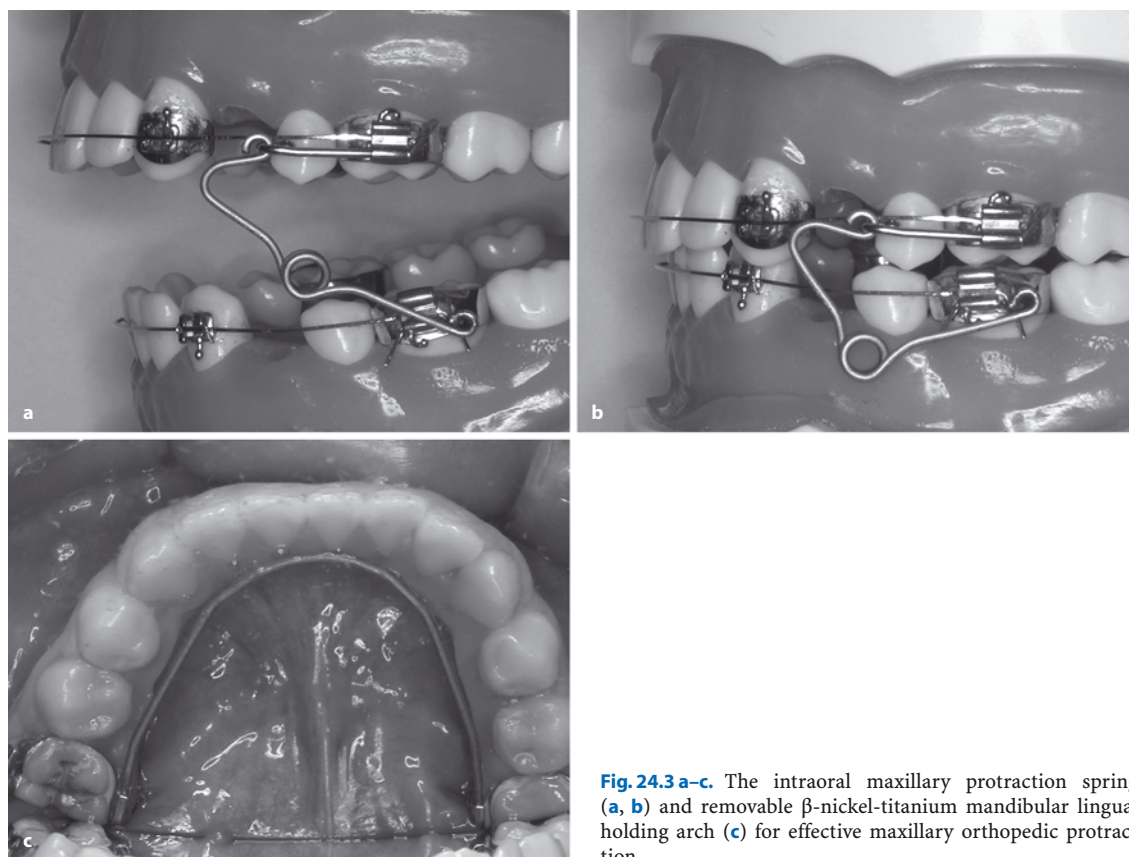


Fig. 24.3 a–c. The intraoral maxillary protraction spring (**a, b**) and removable β -nickel-titanium mandibular lingual holding arch (**c**) for effective maxillary orthopedic protraction

24.2.1.3 Maxillary Protraction Springs for Effective Maxillary Orthopedic Protraction

The maxillary protraction device is a pair of noncompliant, tooth-borne, intraoral maxillary protraction spring (US patent 6273713 B1) [1, 2] (Fig. 24.3). It is a 0.036-inch β -nickel-titanium helix spring. Ball pins are used to mount the spring on the maxillary and mandibular headgear tubes. The spring is activated by the lower jaw movement. It is passively and in 180° when the mandible opens (Fig. 24.3a). While the mandible closes, it is compressed to $100\text{--}120^\circ$ and generated 300–400 gm of horizontal and upward force on the maxilla (Fig. 24.3b). A 0.036-inch β -nickel-titanium mandibular lingual holding arch with built-in lingual crown torque is used for splinting the mandibular dentition as an anchor unit for the protraction (Fig. 24.3c).

24.2.1.4 Treatment Protocol for Effective Maxillary Orthopedic Protraction

The treatment protocol for effective maxillary orthopedic protraction includes 7–9 weeks of Alt-RAMEC and 4 months of maxillary protraction using intraoral maxillary protraction springs. The total treatment protocol is 6 months. The patients are seen every 4 weeks for adjusting or replacing the intraoral maxillary protraction springs when they are distorted or broken. The expander and protraction device are removed at the end of 6th month.

24.2.1.5 Treatment Results and Effects of Effective Maxillary Orthopedic Protraction

The differences between Alt-RAMEC and a single course of rapid maxillary expansion (RME) have been evaluated in a clinical cephalometric study [1]. Twenty-six consecutive cases of unilateral cleft lip and palate patients with hypoplastic maxillae ($\text{SNA} < 82^\circ$)

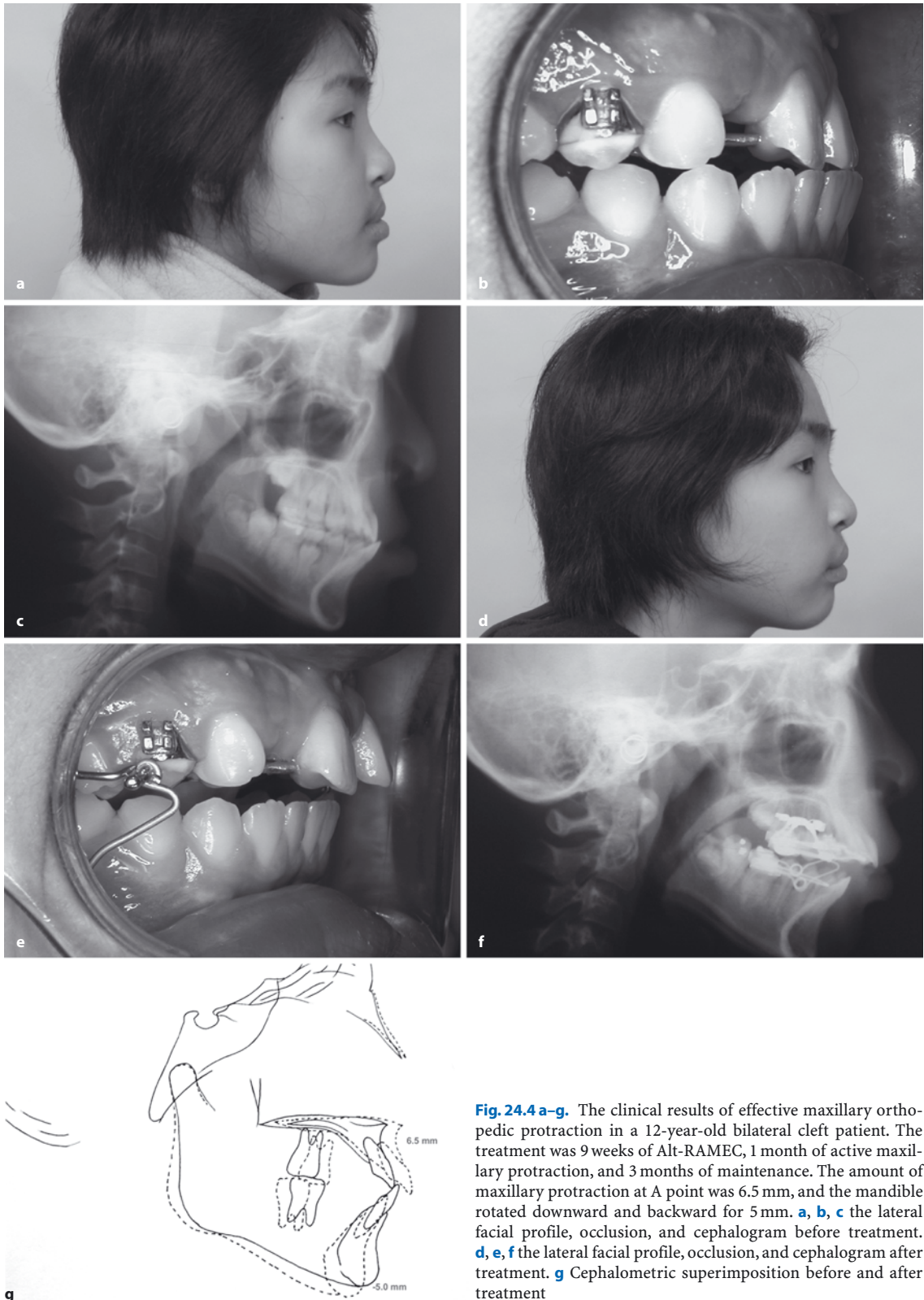


Fig. 24.4 a–g. The clinical results of effective maxillary orthopedic protraction in a 12-year-old bilateral cleft patient. The treatment was 9 weeks of Alt-RAMEC, 1 month of active maxillary protraction, and 3 months of maintenance. The amount of maxillary protraction at A point was 6.5 mm, and the mandible rotated downward and backward for 5 mm. **a, b, c** the lateral facial profile, occlusion, and cephalogram before treatment. **d, e, f** the lateral facial profile, occlusion, and cephalogram after treatment. **g** Cephalometric superimposition before and after treatment

were included for maxillary protraction. Their ages ranged from 9 to 12 years old. The group of RME (7 days, 1 mm/day) was the first 16 consecutive cases. The group of Alt-RAMEC was the next 10 consecutive cases. The expander used in both groups was the double-hinged expander, and the protraction appliance was the intraoral maxillary protraction springs.

The amount of maxillary anterior displacement by the double-hinged expander in the Alt-RAMEC group was 3.0 ± 0.9 mm at A point. This was significantly greater than the 1.6 ± 1.0 mm in the RME group. The amount of maxillary advancement with intraoral protraction springs in the Alt-RAMEC group was 2.9 ± 1.9 mm at A point and was significantly greater than the 0.9 ± 1.1 mm in the RME group. The overall amount of maxillary advancement in the Alt-RAMEC group was 5.8 ± 2.3 mm at A point. The clinical and cephalometric results are shown in Fig. 24.4. The protraction results remained stable without significant relapse after 2 years (5.8 ± 2.3 mm vs. 5.7 ± 3.0 mm).

The protocol of Alt-RAMEC displaces the maxilla anteriorly twice and facilitates maxillary protraction 3 times better than a single course of RME. This indirectly proves that the protocol of Alt-RAMEC disarticulates circummaxillary sutures better than a single course of rapid maxillary expansion. Because the circummaxillary sutures are protracted 3 times faster than usual, the protraction results could be an orthopedic process of “sutural protraction osteogenesis,” which is similar but less vigorous than sutural expansion distraction osteogenesis. The maxillary protraction by using the double-hinged expander, repetitive weekly protocol of Alt-RAMEC, and the intraoral protraction springs is effective with stable results at 2 year follow-up.

To obtain good stability, our clinical experience revealed that the timing for effective maxillary orthopedic protraction is around the onset of puberty (age

11–13). Early treatment is not recommended because of the unpredictable mandibular growth. The onset of puberty could be evaluated by using vertebral bone age on cephalogram [31, 32].

24.3 Orthopedic Management of a Downward Displaced Premaxilla in Bilateral Cleft Patients

In growing bilateral cleft patients, one of the most common premaxillary deformities is a prominent and downward displaced premaxilla with or without wide alveolar clefts (Fig. 24.5a). The vertical discrepancy between the premaxilla and buccal segments results in an unaesthetic “equine appearance,” anterior deep overbite, and also makes the alveolar bone grafting difficult or impossible [33].

In a cephalometric study, Liou et al. inferred that the downward displaced premaxilla is not because of overgrowth of the premaxilla or overeruption of the maxillary incisors [3]. The premaxilla might have been downward distorted in the early age of life after the primary surgical repairs. Several studies indicated that the prominence of premaxilla becomes less pronounced during the growth period and gradually resolves into the adulthood [34–39]. It is recommended that the premaxillary deformities be left untreated until adulthood [40]. However, to leave the deformities unsolved until adulthood may complicate the alveolar bone grafting and impact the young patients psychosocially.

The contemporary surgical interventions of the downward displaced premaxilla in bilateral cleft patients include removal of the primary maxillary incisors in order to improve the appearance and

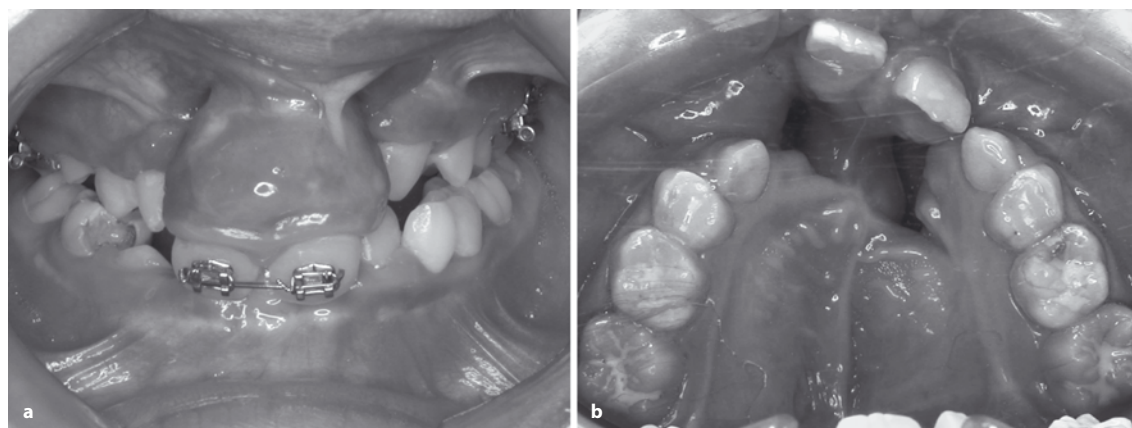


Fig. 24.5 a, b. Two of the most common premaxillary deformities in bilateral cleft lip and palate. **a**, a prominent and downward displaced premaxilla with or without wide alveolar clefts.

b, a laterally displaced premaxilla with a wide alveolar cleft on one side

temporarily reduce the dentoalveolar prominence [38], and surgical reposition of the premaxilla in conjunction with alveolar bone grafting [35, 41–44]. However, the premaxilla in the growing subjects having surgical repositioning grows at a very low rate and becomes retrusive progressively in the older age [38, 45].

24.3.1 Premaxillary Orthopedic Intrusion

A better treatment modality therefore is to reposition the premaxilla and approximate the alveolar cleft nonsurgically and orthopedically so that the growth of premaxilla would not be disturbed and the alveolar bone graft could be performed successfully. A new nonsurgical technique called premaxillary orthopedic intrusion has been developed for correcting the downward displaced premaxilla in growing patients with bilateral cleft [2, 3].

24.3.1.1 Orthodontic Preparation

Before initiating orthopedic intrusion of premaxilla, the maxillary incisors are well aligned orthodontically so that a segment of 0.016 by 0.022 stainless steel arch wire can be placed. The maxillary first permanent molars or the second primary molars are banded and the band is welded with an orthodontic triple tube. The triple tubes are for holding the orthopedic intrusion device. A fan-type rapid maxillary expander, if necessary, could be placed for repositioning the collapsed buccal segments. A transpalatal arch and/or a segment of stainless steel arch wire (0.016 by 0.022) are placed on the buccal teeth for consolidating the buccal segments as an anchor unit for the orthopedic intrusion of premaxilla.

24.3.1.2 Device for Premaxillary Orthopedic Intrusion

No headgear, extraoral device, or surgery is used for the intrusion. The device for orthopedic intrusion of the downward displaced premaxilla is a pair of tooth-borne distraction devices (Fig. 24.6a). The device anchors on the maxillary buccal teeth and delivers an intermittent intrusion force to the premaxilla through the maxillary incisors. The U-shaped connector of the device is inserted into the headgear tube on the molar band and secured with a 0.012-inch ligature wire. The extension arm of the device is secured with 0.012-inch ligature wires onto the segmental arch wire on the central incisors (Fig. 24.6 b–e).

On both sides, the devices are activated 0.3 mm per day until the premaxilla has been upward repositioned to the desired position. The devices are then maintained for another 3 months before their removal. After removal of the intrusion devices, an orthodontic arch wire is placed for further maintenance, and alveolar bone grafting is then performed within 3 months.

24.3.1.3 Treatment Results of Premaxillary Orthopedic Intrusion

The treatment results have been evaluated in a cephalometric study on 10 consecutive cases [3]. The orthopedic intrusion of premaxilla was completed within 4 weeks in all the patients. After the intrusion, the occlusal planes and gingival lines of the premaxilla and buccal segments were leveled clinically. The facial appearance during resting and smiling improved and had no “equine appearance.” The clinical results are shown in Fig. 24.7.

The cephalometric study revealed that there was no significant vertical movement of the maxillary buccal segments and the premaxilla was significantly intruded (Fig. 24.7d). The nasal bone was also significantly brought forward and upward at the nasal tip, but its amount was significantly less than the orthopedic intrusion of the premaxilla.

The correction of the premaxillary deformity mainly came from three aspects, orthopedic intrusion of the premaxilla, dental intrusion of the maxillary incisors, and shortening of the premaxillary dentoalveolar height. The orthopedic intrusion of the premaxilla (3.0 mm) and dental intrusion of the maxillary incisors (3.5 mm) gave a total amount of 6.5 mm upward repositioning at the tip of the maxillary incisor. The shortening of the premaxillary dentoalveolar height (3.5 mm) could be due to the dental intrusion of the maxillary incisors, which was expected when a tooth-borne distraction device was used. The correction was 46% (3.0/6.5) orthopedic effect and 54% (3.0/6.5) dentoalveolar effect.

The orthopedic intrusion of the premaxilla remains relatively stable in the 1-year observation after treatment. However, the dental intrusion of maxillary incisors seems unstable. The dental relapse of the maxillary incisors tends to elongate the premaxillary dentoalveolar height and compromise the result of premaxillary orthopedic intrusion. For this reason, it is recommended to maintain the dental intrusion with an orthodontic arch wire and perform alveolar bone grafting shortly after removal of the intrusion devices (Fig. 24.7c).

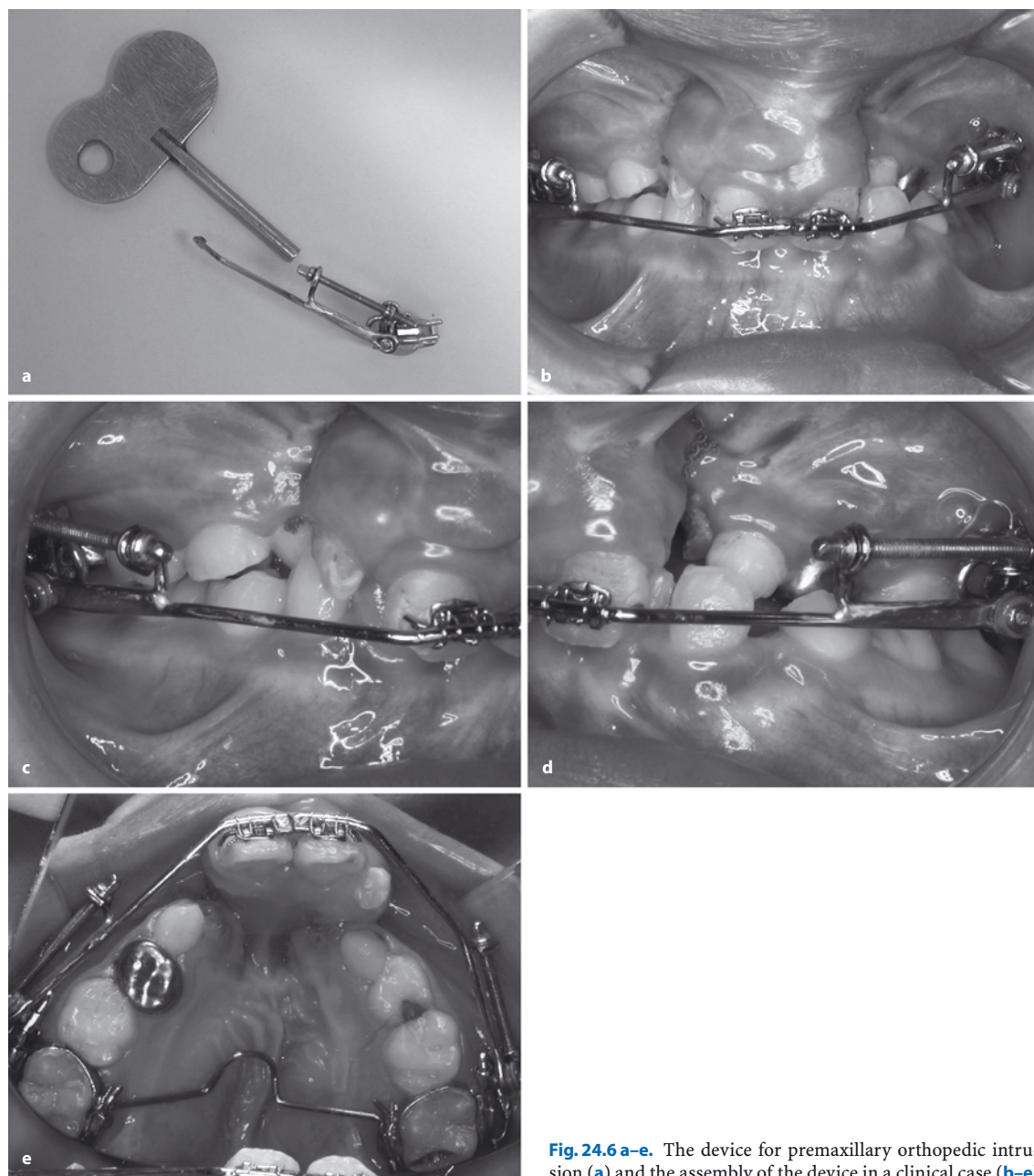


Fig. 24.6 a–e. The device for premaxillary orthopedic intrusion (**a**) and the assembly of the device in a clinical case (**b–e**)

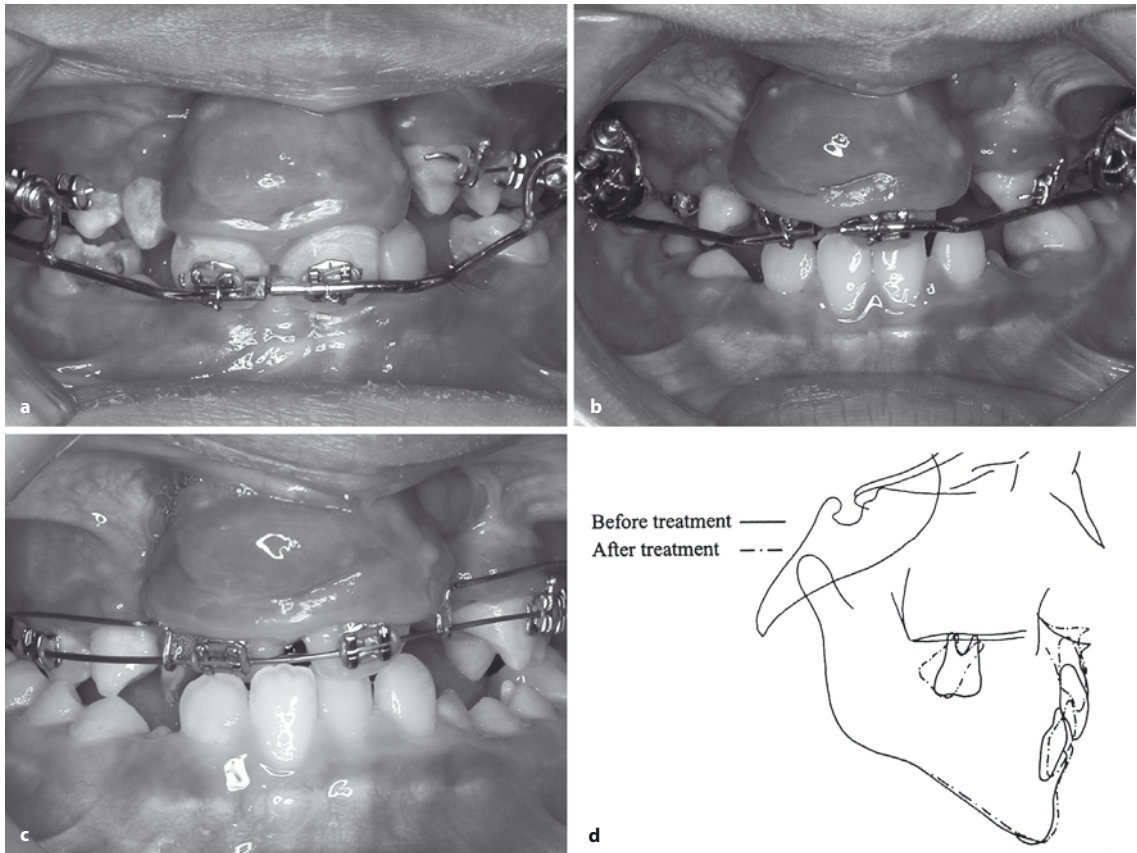


Fig. 24.7 a–d. The premaxillary orthopedic intrusion. The equine appearance of the premaxilla prior treatment (**a**), after 1 month of orthopedic intrusion (**b**), 5 months after the ortho-

pedic intrusion (**c**), and cephalometric superimposition before and after premaxillary orthopedic intrusion (**d**)

24.3.1.4 Mechanisms of Premaxillary Orthopedic Intrusion

The possible events that may occur during orthopedic intrusion of premaxilla include mechanical upward displacement of the premaxilla and vomero-nasal septum complex, sutural contraction osteogenesis [46] and/or osteolysis [47] in the vomero-premaxillary suture, or bending/remodeling of the vomero-nasal septum complex:

1. Mechanical upward displacement without bending/remodeling of the vomero-nasal septum complex and no sutural contraction osteogenesis/osteolysis in the vomero-premaxillary suture
2. Sutural contraction osteogenesis/osteolysis in the vomero-premaxillary suture without bending/remodeling/mechanical upward displacement of the vomero-nasal septum complex

3. Bending/remodeling without mechanical upward displacement of the vomero-nasal septum complex and no sutural contraction osteogenesis/osteolysis in the vomero-premaxillary suture
4. Any combinations of 1, 2, or 3.

By the analysis on the lateral and posteroanterior cephalometric radiographs, it was implied what occurred during the orthopedic intrusion of premaxilla was mostly similar to the sutural contraction osteogenesis/osteolysis in the vomero-premaxillary suture combined with slightly mechanical upward displacement of the vomero-nasal septum complex and nasal bones (Fig. 24.8).

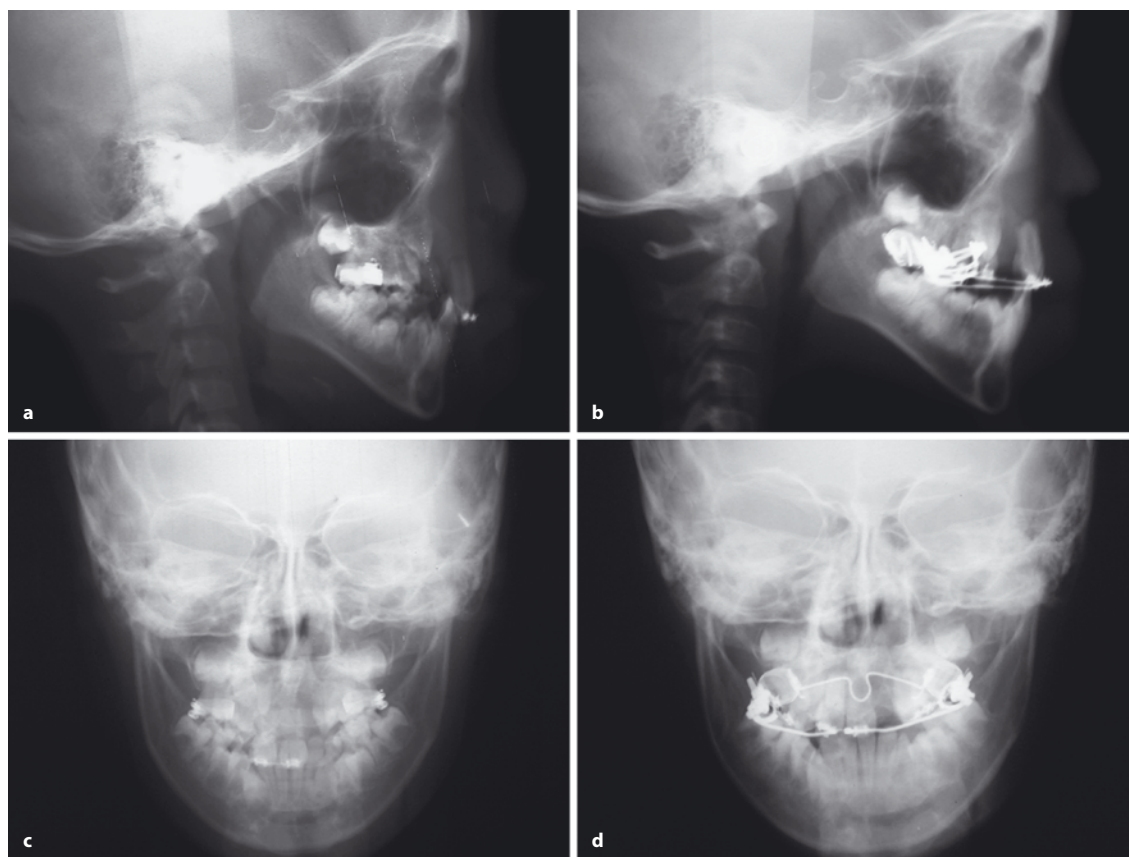


Fig. 24.8 a–d. The cephalometric radiographs of the same patient in Fig. 24.7. **a, b** the lateral cephalogram before and after premaxillary orthopedic intrusion. **c, d** The posteroanterior

cephalogram before and after premaxillary orthopedic intrusion. Note the premaxilla and incisors were intruded without further deviation of the vomer bone and nasal septum

24.4 Orthopedic Management of a Laterally Displaced Premaxilla in Bilateral Cleft Patients

In growing bilateral cleft patients, another common premaxillary deformity is a laterally displaced premaxilla with a wide alveolar cleft on one side (Fig. 24.5b). The laterally displaced premaxilla results in an unaesthetic appearance with a skewed premaxilla and dental midline, and uneven width of the alveolar cleft beside the premaxilla.

The alveolar bone grafting and closure of oronasal fistula could be difficult without surgical repositioning the laterally displaced premaxilla. However, the premaxilla in growing subjects having surgical repositioning grows at a very low rate and becomes retrusive progressively as they age [38, 45].

24.4.1 Premaxillary Orthopedic Medial Repositioning

A better treatment modality therefore is to reposition the premaxilla and approximate the alveolar cleft nonsurgically and orthopedically so that the growth of premaxilla would not be disturbed and the alveolar bone graft could be performed successfully. A new nonsurgical technique called premaxillary orthopedic medial repositioning has been developed for correcting the laterally displaced premaxilla in growing patients with bilateral cleft [2].

24.4.1.1 Orthodontic Preparation

Similar to the clinical procedures for the premaxillary orthopedic intrusion, the entire maxillary dentition is aligned orthodontically so that a 0.016 by 0.022 stainless steel arch wire could be placed before the orthopedic repositioning. The maxillary first permanent molars are banded and the band is welded with an orthodontic triple tube. For the anchorage, a transpalatal arch is placed for consolidating both of the buccal segments as an anchor unit for the orthopedic repositioning of premaxilla.

24.4.1.2 Device for Premaxillary Orthopedic Medial Repositioning

No headgear, extraoral device, or surgery is used for the repositioning. The device for orthopedic medial repositioning is an intraoral tooth-borne distraction device (Fig. 24.9). It consists of a body, a screw, and an extension arm. There are two bolts on the body for holding the screw. The fixed bolt is attached on the front of the body for holding the head of the screw. The sliding bolt rides and slides on the sliding bar of the body for holding the tail of the screw. The U-shaped connector at distal end of the body is inserted into the headgear tube on the molar band and secured with a 0.012-in ligature wire. The extension arm is adjusted to hook distally to the maxillary incisors so that the premaxilla is being pushed toward the midline when the device is activated. The arch wire is a track guiding the premaxilla to slide in a linear curvature direction during the orthopedic repositioning.

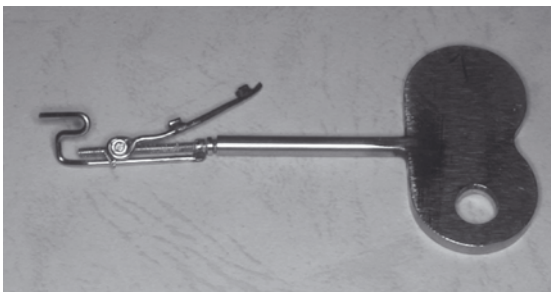


Fig. 24.9. The device for premaxillary orthopedic medial repositioning

The screw is activated 0.3 mm per day until the maxillary dental midline has been overcorrected for 2–3 mm. The device is then maintained for another 3 months before its removal. After the removal, orthodontic elastics or ligature wire is placed for further maintenance before the next treatment on the alveolar cleft.

24.4.1.3 Treatment Results of Premaxillary Orthopedic Medial Repositioning

The treatment results have been evaluated in a cephalometric study on four consecutive cases [2]. The clinical results are shown in Fig. 24.10. The age of the patients were 9 to 12 years old at the time of treatment. Before the treatment, the angular deviation of premaxilla and nasal septum ranged from 15° to 25°, and the linear deviation of dental midline ranged from 5 mm to 8 mm as measured on the posteroanterior cephalograms.

The orthopedic medial repositioning of premaxilla was completed within 2 to 3 weeks in all the patients. All the patients tolerated the device well and no pain was reported by any of the patients. The maxillary dental midline was overcorrected for 2–3 mm. The overall correction of the dental midline ranged from 7 mm to 11 mm. During the maintenance period (3 months), the premaxilla and maxillary dental midline gradually moved back toward the mandibular dental midline. The smile facial appearance was improved and the premaxilla and dental midline were repositioned on the midline.

24.4.1.4 Mechanisms of Premaxillary Orthopedic Medial Repositioning

The posteroanterior cephalometric and occlusal radiographs before and 3 months after the repositioning were evaluated. It was revealed that both the deviated nasal septum and premaxilla were all straightened (Fig. 24.11). The straightening of the nasal septum and premaxilla could be due to bone bending and remodeling of the vomer bone. The angular correction of the premaxilla and nasal septum ranged from 10° to 20°, and the linear correction of the premaxilla ranged from 5 mm to 10 mm, as measured on the radiographs.

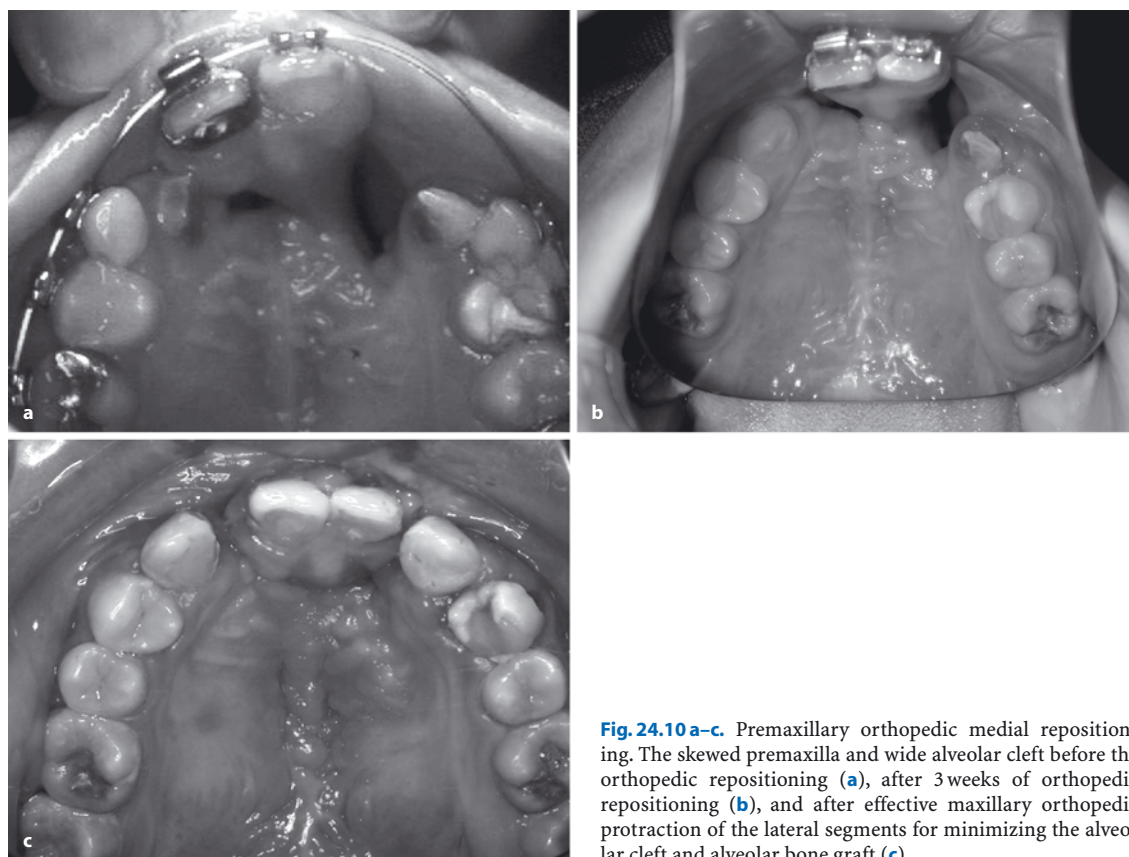


Fig. 24.10 a–c. Premaxillary orthopedic medial repositioning. The skewed premaxilla and wide alveolar cleft before the orthopedic repositioning (**a**), after 3 weeks of orthopedic repositioning (**b**), and after effective maxillary orthopedic protraction of the lateral segments for minimizing the alveolar cleft and alveolar bone graft (**c**)

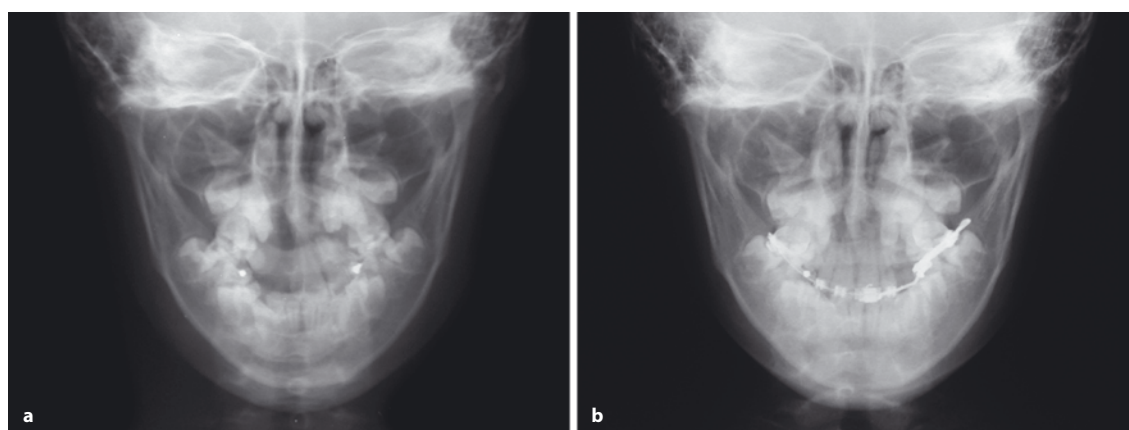


Fig. 24.11 a,b. The posteroanterior cephalograms reveal straightening of the vomer bone after premaxillary orthopedic medial repositioning (**a**, before; **b**, after medial repositioning)

24.5 Managements of a Wide Alveolar Cleft and Fistula

Although premaxillary orthopedic intrusion or orthopedic medial repositioning is able to correct the downward or laterally displaced premaxilla, the alveolar cleft might be still be too wide to be bone grafted. Using oral mucosa at a distant site, such as rotation advancement flap of buccal mucosa or tongue flap [48], could close a wide alveolar cleft. By such approach, the alveolar cleft is not bone grafted and the maxillary segments are not approximated. Neither the buccal mucosa nor the tongue mucosa is a good substitute for the attached gingiva. Attached gingiva is essential for the subsequent tooth eruption, orthodontic tooth movement, or dental prostheses after alveolar bone grafting and closure of fistula [48, 49].

Another approach is to approximate the alveolar cleft through LeFort I osteotomy and advancement so that both the soft and bony tissue could be well approximated and bone grafted [50]. However, LeFort I advancement might disturb the growth of maxilla in young patients [51].

24.5.1 Protocol for Approximating a Wide Alveolar Cleft

We have developed a protocol for minimizing or approximating a wide alveolar cleft [2].

1. For an alveolar cleft less than a tooth width, effective maxillary orthopedic protraction of the maxillary lateral segments and alveolar bone grafting is the treatment of choice.
2. For an alveolar cleft wider than a tooth width, interdental distraction osteogenesis [16] and alveolar bone grafting or gingivoperiosteoplasty is the treatment of choice.

24.5.2 Minimize Alveolar Cleft by Effective Maxillary Orthopedic Protraction

The lateral segments of maxilla are orthopedically protracted toward the premaxilla to minimize the alveolar cleft. The rationales and mechanisms are similar to entire maxillary protraction by using effective maxillary orthopedic protraction. The total treatment protocol is 6 months, including 8–9 weeks of Alt-RAMEC followed by 4 months of orthopedic protraction of the lateral segments of maxilla. The expander is a double-hinged rapid maxillary expander, and the protraction appliance is the intraoral maxillary protraction springs.

24.5.2.1 Clinical Procedures for Effective Maxillary Orthopedic Protraction of Lateral Segments of Maxilla

Maxillary molars and primary canines (or first premolars) are banded and maxillary impression is taken for fabricating the double-hinged rapid maxillary expander. The expander is oriented perpendicular to the central incisors. The expander anchors only on lateral segments of maxilla, and there is no extension arm on the premaxilla. The inner surfaces of the bands are sandblasted before cementation. One day after the cementation, the double-hinged expander is activated according to the protocol of Alt-RAMEC and then is left in place for orthopedic protraction of the lateral segments of maxilla. A 0.036-inch β -nickel-titanium mandibular lingual holding arch with built in lingual crown torque is used for splinting the mandibular dentition as an anchor unit for the orthopedic protraction. Patients are seen every 4 weeks for adjusting or replacing the intraoral maxillary protraction springs when they are distorted or broken. The expander and the springs are then removed at the end of the 6th month.

24.5.2.2 Treatment Results

Three of the four patients who had premaxillary orthopedic medial repositioning received the treatment. The treatment results were evaluated clinically and cephalometrically [2]. All of the alveolar clefts were successfully minimized, and the gingival and mucosal tissue beside the alveolar clefts was approximated after the orthopedic protraction (Fig. 24.12). All of the patients received orthodontic treatment after removal of the expander and protraction springs. The alveolar bone grafting was then performed shortly after the maxillary dentition was well aligned.

On the cephalometric evaluation, the lateral segments of maxilla were protracted for 2–3 mm. The buccal teeth on the maxillary lateral segments were also moved forward for 2–3 mm. The alveolar clefts were all minimized to 2–3 mm as measured on the radiographs. No retrusion or retraction of the premaxillae was observed. The overjet at anterior teeth was maintained positive through out the treatment.

The maxillary lateral segment orthopedic protraction by Alt-RAMEC is an effective nonsurgical technique for minimizing the alveolar cleft in patients with BCLP. This technique is much less invasive relative to the surgical repositioning of premaxilla. The treatment effects are both the orthopedic protraction of the maxillary lateral segments and forward movement of the buccal teeth.

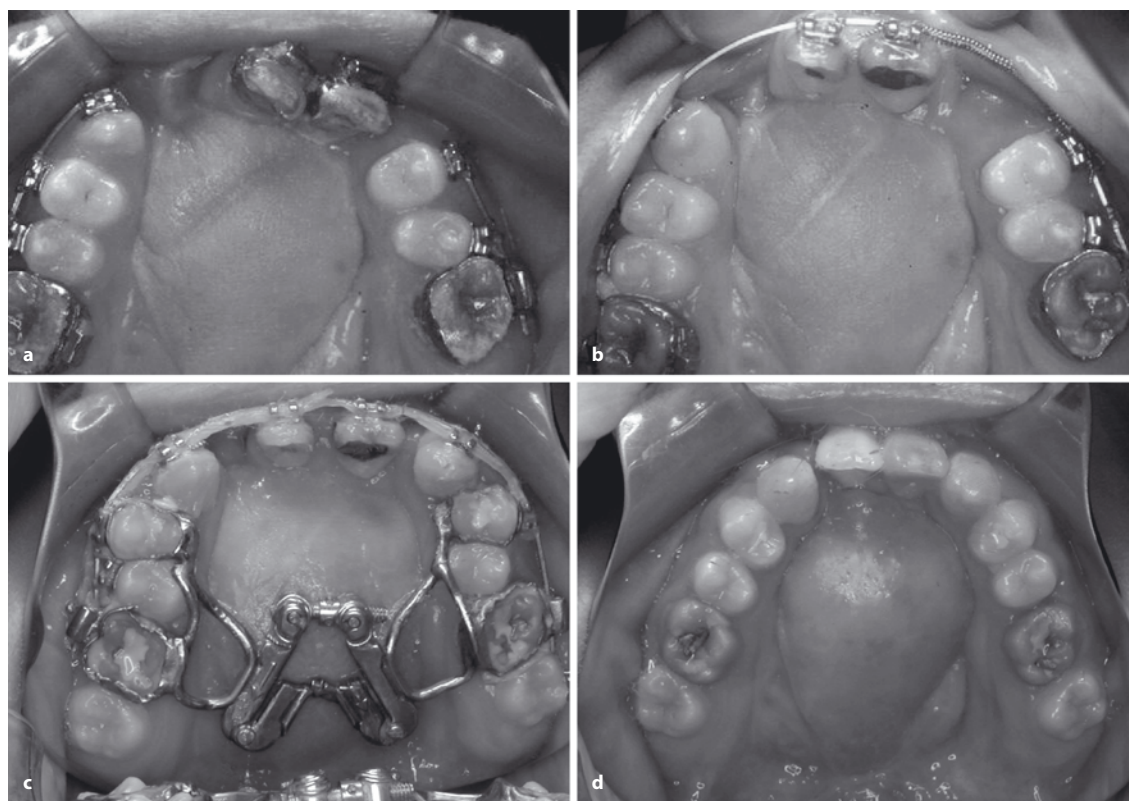


Fig. 24.12 a–d. Minimize alveolar cleft less than a tooth width. The skewed premaxilla and alveolar cleft before the orthopedic repositioning (**a**), after 3 weeks of orthopedic medial repositioning (**b**), during effective maxillary orthopedic protraction of the lateral segments (**c**), and after the orthopedic protraction and alveolar bone graft (**d**)

24.5.3 Interdental Distraction Osteogenesis for Approximating Alveolar Cleft Wider than a Tooth Width

The closure of an alveolar cleft wider than a tooth width is a challenge not only because of the difficulty in complete closure by using local attached gingiva but also the great volume of bone grafting. Interdental distraction osteogenesis (IDO) has been proposed to approximate wide alveolar clefts and to achieve bony union across the clefts without bone grafting, either in unilateral or bilateral cleft [4]. It is a technique of dentoalveolar lengthening through soft callus distraction osteogenesis. The dentoalveolus is osteotomized interdentially, and then transported toward the cleft so that the cleft could be approximated by growing local alveolus and attached gingiva at a distant site to the cleft (Fig. 24.13.). Its advantages include eliminating the need for extensive alveolar bone grafting, creating dental space (the regenerate alveolus and attached gingiva) for relieving dental crowding, as well as avoiding worsening the velopharyngeal insufficiency.

24.5.3.1 Presurgical Orthodontic Preparations

All the maxillary teeth are bonded and banded and then they are well aligned so that a 0.016 by 0.022 (or thicker) stainless steel arch wire could be placed. A transpalatal arch could be placed for consolidating buccal segments as one unit when it is necessary. One of the purposes of orthodontic tooth alignment before interdental distraction osteogenesis is to well align all the maxillary teeth so that the osteotomized dentoalveolus can slide along the arch wire during distraction. Besides the distraction device, the stainless steel arch wire also is a guiding track for interdental distraction osteogenesis. The distraction device has a linear track, but the arch wire guides the osteotomized dentoalveolus to be distracted along the arch wire in a linear curvature direction (Fig. 24.13b).

An interdental space is opened orthodontically at the selected distraction site by using an open coil spring or similar mechanisms. This is to separate the

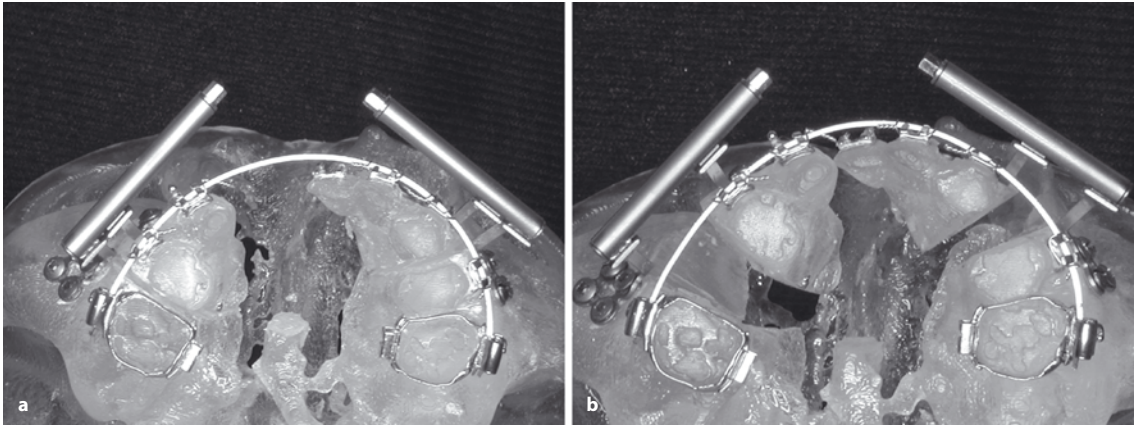


Fig. 24.13 a,b. Model illustrations of interdental distraction osteogenesis. The dentoalveolus is osteotomized interdentally (**a**) and transported toward the cleft by the distractor (**b**). The

orthodontic arch wire also is a guiding track. The osteotomized dentoalveolus slides on the arch wire in a linear curvature direction

dental roots and increase the thickness of the interdental alveolar bone so that interdental osteotomy could be performed without damaging or stripping the adjacent dental roots.

24.5.3.2 Interdental Distraction Site

Although the selection of distraction site varies regarding different clinical situations of the cleft, there are certain guidelines.

1. The osteotomized segment of dentoalveolus to be distracted should have adequate blood supply from adjacent gingival or oral mucosa, and adequate bone volume to sustain a successful distraction osteogenesis. A width of at least two teeth is recommended.
2. The interdental distraction site should be wide enough for interdental osteotomy and to avoid damaging or stripping the adjacent dental roots during interdental osteotomy. At least 3 mm is recommended.
3. At least 1.0 mm thickness of interdental alveolar bone should be preserved beside the adjacent dental roots after interdental osteotomy. Inadequate thickness could result in severe periodontal problems, such as exposure of the adjacent dental roots and insufficient volume and height of the regenerate.
4. The interdental distraction site should have enough attached gingiva for a primary closure. Exposure of the osteotomy site would result in wound infection, sequestrae, and distraction failure.

24.5.3.3 Surgical Procedures

The surgical procedures are performed under nasotracheal anesthesia. A horizontal intraoral incision is made along the buccal vestibule of the maxilla. Superior based mucoperiosteal flaps are reflected, exposing the site of horizontal maxillary osteotomy on the buccal side. A vertical mucoperiosteal tunnel extending upward from the interdental attached gingiva to the horizontal incision is made to expose the site of vertical interdental osteotomy. Another small incision, on the palatal side, is made inside the gingival sulcus, exposing the site of interdental osteotomy. Care is taken to avoid reflecting or stripping all of the palatal mucosa.

By estimating on the radiographs, the anatomic position of the dental roots, sites of interdental and horizontal maxillary osteotomies, and permanent tooth buds are marked on the alveolar bone with surgical marking pens. Complete horizontal osteotomy is performed with a cutting saw, 3–5 mm away from the dental root apex and tooth buds. Complete interdental osteotomy is then performed with a small round bur and followed by a thin osteotome, cutting through the buccal and palatal cortical plates respectively. As the cutting round bur encounters the cancellous bone, the thin osteotome is then used for cutting carefully through the interdental cancellous bone buccolingually. After the horizontal maxillary and interdental osteotomies, the distal segment of the osteotomized dental arch is completely mobile. The orthodontic arch wire holds the osteotomized segment in place after osteotomies and prevents medial collapse of the segment when the distraction device is placed.

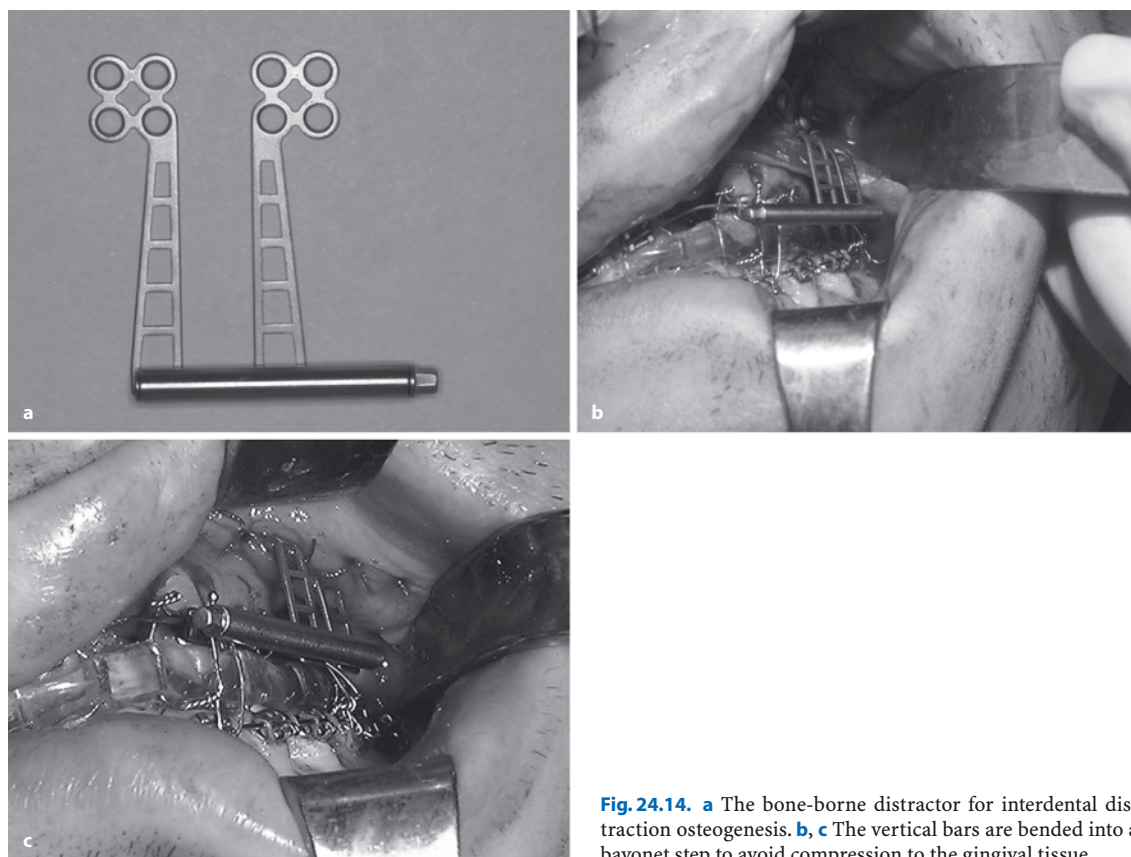


Fig. 24.14. **a** The bone-borne distractor for interdental distraction osteogenesis. **b, c** The vertical bars are bended into a bayonet step to avoid compression to the gingival tissue

The distraction device is a bone-borne device (Martin & KLS, USA) (Fig. 24.14a). The vertical bars of the bone-borne distractor are bended into the desired shape to avoid any possible compression to the gingival adjacent to the vertical osteotomy sites (Fig. 24.14b). The direction of the distractor needs careful adjustment to ensure a correct vector for bony movement. It also needs great care to avoid any compression of the distractor to the buccal mucosa. The distractor is then fixed to maxilla with unicortical screws. The incisions are irrigated and closed with absorbable sutures (Fig. 24.14c). A very important point in wound closure is closing the attached gingiva over the interdental osteotomy sites to avoid any possible wound separation complicated with dental root exposure in the vertical osteotomy site.

24.5.3.4 Distraction Protocol

The latency period is 7 days. This allows enough time for the formation of soft callus and primary healing of the attached gingiva and oral mucosa. The soft tissue wound healing is critical for the distraction results. Soft tissue wound dehiscence during distraction could result in severe bone resorption, gingival recession, and exposure of the dental root. The distraction device is activated 1 mm/day until both ends of the alveolar cleft has been approximated. After completion of interdental distraction, the device is left in place for 3 months for maintenance and postdistraction orthodontic tooth movement through regenerate.

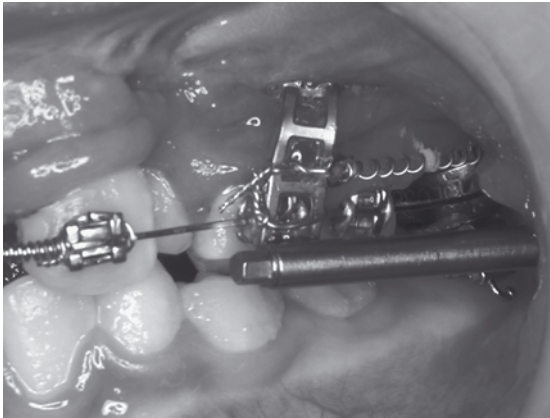


Fig. 24.15. Postdistraction orthodontic tooth movement. Orthodontic nickel-titanium coil spring is attached between the maxillary first molar and the vertical bar of the distractor for moving the first molar into the regenerate

24.5.3.5 Postdistraction Maintenance and Orthodontic Tooth Movement Through the Regenerate

Two weeks after completion of the interdental distraction, the tooth/teeth right adjacent to the interdental distraction, either the mesial or distal one or both, are moved orthodontically into the regenerate. This is to eliminate the interdental space created by distraction, or to utilize the created space for relieving dental crowding.

Orthodontic elastic chains or nickel-titanium coil springs are used for moving tooth/teeth into the regenerate. They are placed between the moved tooth/teeth and the vertical bars of the bone-borne distraction device (Fig. 24.15). The bone-borne distraction device holds the distracted segment through several bone screws, while the tooth/teeth at the distracted segment are being moved through the regenerate without shortening the width of the regenerate. The bone-borne distraction device is left in place for not only maintenance but also is an anchor for postdistraction orthodontic tooth movement. This is similar to the implant orthodontics that uses bone screws or plates as anchorage for orthodontic tooth movement [52]. After the tooth/teeth have moved into the regenerate, the adjacent tooth/teeth are then moved subsequently into the left space. The postdistraction orthodontic tooth movement usually can be completed in 3 months because the regenerate is still soft.

24.5.3.6 Postdistraction Alveolar Bone Grafting or Gingivoperiosteoplasty

After the interdental distraction osteogenesis and postdistraction orthodontic tooth movement, either conventional alveolar bone grafting or gingivoperiosteoplasty can be performed as the final treatment procedure for closing the wide alveolar cleft. Soft tissue approximation of the alveolar cleft does not mean bony approximation of the alveolar cleft. Occlusal or periapical film is needed for revealing the remaining width of the bony alveolar cleft.

1. The alveolar bone grafting is performed in cases where the bony gap between the alveolar cleft is still wider than 2 mm after interdental distraction osteogenesis.
2. The gingivoperiosteoplasty is performed in cases where the bony gap between the alveolar cleft is less than 2 mm after the interdental distraction osteogenesis.

24.5.3.7 Treatment Results

Twenty-one patients, including 13 unilateral and 8 bilateral clefts who had interdental distraction osteogenesis for approximating their wide alveolar cleft in 1998–2002 were reviewed for their treatment results (Fig. 24.16). The follow-up period was 4–5 years. There were 29 alveolar clefts and the average width was 10 mm.

The distracted segments of the dental arches were moved almost bodily toward the cleft as revealed on the panoramic and cephalometric radiographs. The average amount of distraction, as measured on the radiographs, was 12 mm with a range of 10 mm to 20 mm. The relapse was 0.5 mm in the first 3 months after removal of distraction device.

Clinically, 28 out of 29 alveolar clefts were approximated completely by soft tissue contact after interdental distraction osteogenesis. Thirteen out of the 29 alveolar cleft were successfully bone grafted, and 16 out of the 29 alveolar cleft had gingivoperiosteoplasty that resulted in bony union across alveolar cleft. The 4- to 5-year follow-up revealed the treatment results were stable.

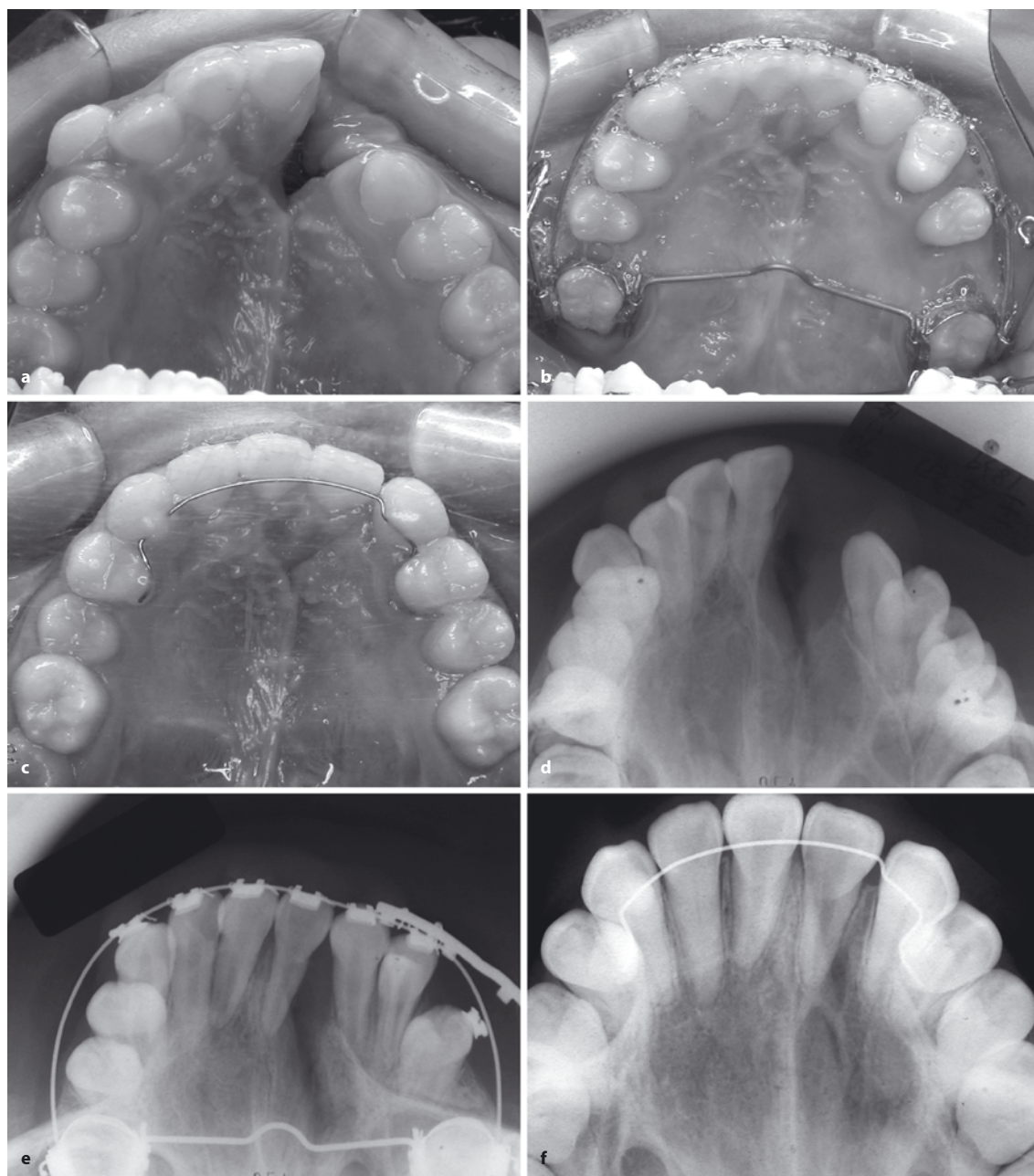


Fig. 24.16 a–f. Approximation of a wide alveolar cleft by interdental distraction osteogenesis in a 12-years-old unilateral cleft patient. The alveolar cleft is bone grafted after the distraction.

a, b, c The clinical photos before, immediately after distraction, and 2 years after distraction. **d, e, f** The occlusal films before, immediately after distraction, and 2 years after distraction

24.6 Summary

In this chapter, we introduced three new orthodontic and orthopedic techniques and one surgical distraction osteogenesis for the management of maxillary deformities in growing unilateral and bilateral cleft patients. These techniques are the effective maxillary orthopedic protraction for correcting a hypoplastic maxilla and minimizing alveolar cleft, premaxillary orthopedic intrusion for correcting a downward displaced premaxilla, premaxillary orthopedic medial repositioning for correcting a lateral displaced premaxilla, and interdental distraction osteogenesis for approximating a wide alveolar cleft. These techniques utilize principles of distraction osteogenesis. The orthopedic approaches could be a form of sutural expansion or protraction osteogenesis, and their treatment effects are mostly orthopedic and partly orthodontic. The interdental distraction is a form of callus distraction osteogenesis. The clinical and radiographic evaluations have revealed their successful applications for solving maxillary deformities in growing cleft patients.

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25 Remodeling the Craniofacial Skeleton by Distraction Osteogenesis – The Mandible

FERNANDO MOLINA

Distraction osteogenesis is becoming the treatment of choice for the surgical correction of mandibular hypoplasias. The development of the technique represents a significant advancement in the field of craniofacial surgery. Deficiencies in the growth of the mandible may result from condylar fractures suffered at an early age affecting growth centers or severe sepsis with secondary condyle resorption resulting with temporomandibular joint ankylosis and micrognathia. Congenital deformities such as Goldenhar's syndrome, Nager's syndrome, craniofacial scoliosis, Pierre Robin sequence, and hemifacial microsomia, may present with mandibular hypoplasia of varying severity [1–4].

Mandibular distraction is a technique less invasive and time-intensive and has a significantly decreased morbidity rate compared with traditional methods of mandibular reconstruction. A surgeon is now able to generate new bone in patients with a bilateral mandibular body deficiency, in a severe hypoplastic ascending ramus, or to reconstruct a new condyle when it is missing in the ankylotic patient. The technique also provides the added benefit of expanding the overlying soft tissues. It probably represents the first tissue engineering surgical technique applied into the craniofacial field.

Bone lengthening or distraction osteogenesis using osteotomies and circumferential gradual distraction was described by Ilizarov [5, 6] to align fractured segments of long bones and later to elongate these bones without a bone graft.

Although the technique initially was developed to lengthen the long bones, in 1973, Snyder reported mandibular lengthening in a canine model using an extraoral device [7]. A similar report on two dogs using an intraoral device followed from Italy [8]. More recently, Karp and McCarthy reported membranous bone lengthening using external devices and that cortical bone formed in the expanded area of the mandible [9]. Histologic examination of the zone revealed a highly organized biologic process.

Since the beginning of the 1990s successful mandibular distraction has been performed on a multitude of patients around the world [10–16]. Our group has been performing mandibular distraction since 1990 using external corticotomy and flexible uni- and bidirectional external devices to achieve simultaneous skeletal and soft tissue correction with minimal surgery [17–21]. The technique has proven to be a major advance in the treatment of a variety of craniofacial deformities. In the midface animal models with or without osteotomies [22, 23] maxillary distraction led to the first clinical trial of this concept in patients with cleft lip and palate [24–26] and craniosynostosis [27–32]. We will include also our experience with distraction osteogenesis in craniosynostosis to achieve postero–anterior movement of the whole facial mass and a combined zygoma-malar rotation with the aim of establishing a proper dental occlusion, of increasing orbital capacity and of improving craniofacial form with a well-balanced proportion.

25.1 Clinical Applications of Distraction Osteogenesis

25.1.1 Hemifacial Microsomia

Facial asymmetry and microtia are the most important clinical findings in a patient. Deviation of the chin to the affected side, hypoplasia of the soft tissues, and associated disorders of other anatomic structures such as the maxilla, the zygoma, and the muscles of mastication are present in a wide variability. The hypoplasia affects the gonial angle (Grade I) in less severe cases and the angle and the ascending ramus in others (Grade IIA and IIB) and show a complete absence of ramus and the condyle in more severe cases (Grade III), [33].

The operative technique begins with a 3 to 5 cm vestibular incision made under general anesthesia.

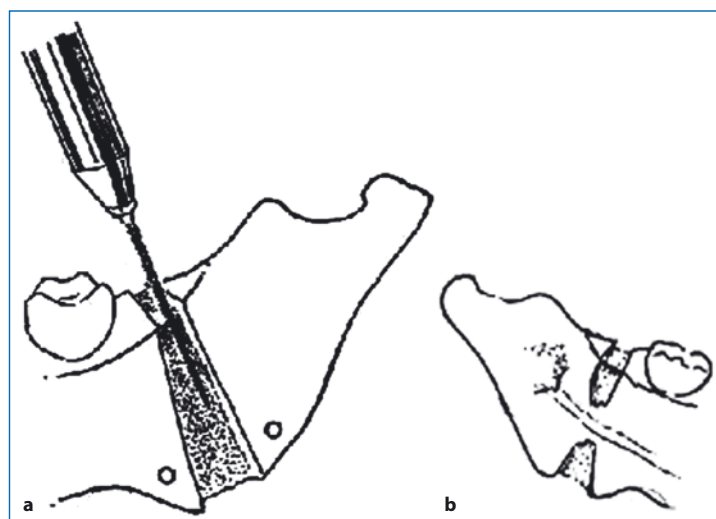


Fig. 25.1 a, b. Side-cutting burr and the external-extended corticotomy from the free mandibular border to the gonial angle. The corticotomy is interrupted as soon as the cancellous bone is visible. **b** At the lingual cortex, the bone cut is incompletely, preserving 6–7 mm of the internal cortical flayer at the site of the neurovascular bundle

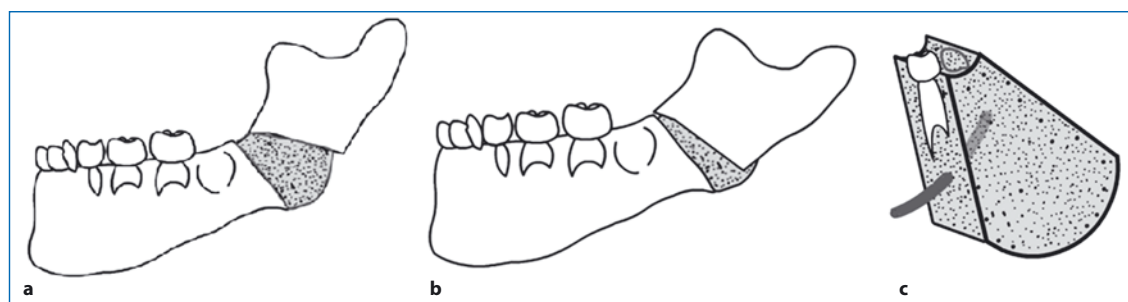


Fig. 25.2. a Location of the corticotomy in a Grade II-A hemifacial microsomia. **b** Using an oblique vector, the new bone formation production is larger at the angle and minor at the alveolar ridge, also at the initial portion of the ascending ramus.

c Tridimensional diagram of the regenerate bone. The new volumetric bone formation corrects the height, the length, and position of the mandible. The tooth buds and neurovascular elements have been preserved

The periosteum is elevated to expose the gonial angle, with a side-cutting burr; the corticotomy is done on the lateral aspect of the mandible, including all the cortex to the cancellous layer. It extends obliquely from the free mandibular border to the gonial angle. Then, the corticotomy is extensively extended inferiorly around the edge of the angle. In fact, 6–7 mm of the internal cortical layer remains intact, protecting the neurovascular bundle, and in this manner the whole cancellous layer, the circulation, and innervation of the bone are preserved (Fig. 25.1 a, b).

Afterwards, a flexible unidirectional external device (KLS-Martin, Germany) is inserted. Two titanium pins are introduced percutaneously through the whole thickness of the mandible, 4–5 mm in front and behind the corticotomy. The pins should be parallel to each other to facilitate the fixation of the distraction device. In this moment, we recommend an “intraoperative test” of accuracy: activate the device 5–6 mm,

and observe the changes in the corticotomy. The surgeon must obtain the opening of the corticotomy segments without difficulty and assess that the elongation system works properly. If the surgeon identifies the presence of cortical bridges, the lengthening mechanisms will not work and the corticotomy must be revised.

The advantage of combining the use of an external corticotomy and a flexible device is the ability to obtain a 3D correction of height, length, and position of the mandible. In fact, different zones of bone resistances are encountered, minor of the angle but major resistance at the alveolar ridge, that will be elongated differently under a perpendicular distraction vector (Fig. 25.2 a–c).

The area of new bone formation produces a better shape of the mandibular angle and modifies the ascending ramus position with a simultaneous better relationship of the condyle and the glenoid fossae.

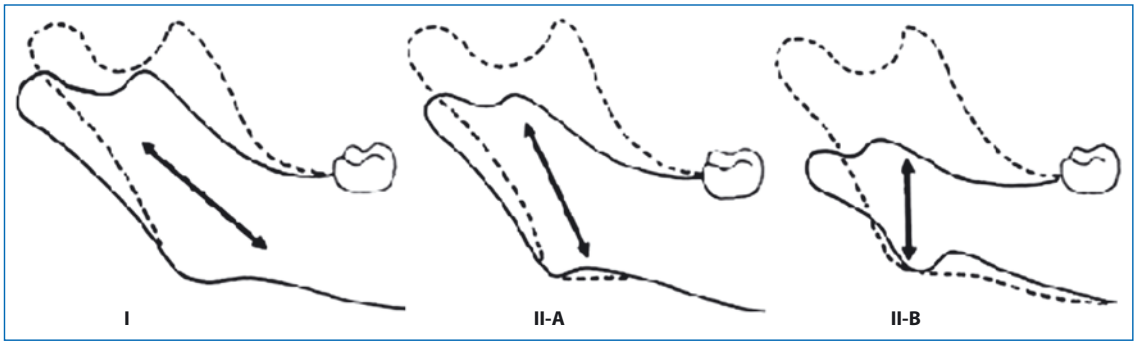


Fig. 25.3. Distraction vectors are the key to obtaining excellent results. Basically, three distraction vectors are used to correct a mandibular hypoplasia in hemifacial microsomia. The oblique

vector is used for Grade Pruzansky I and the vertical vector is used to recreate the ascending ramus in the Grade Pruzansky II-B

The initial condyle lateral position moves into a more central one that is more similar with the contralateral normal one. All of these findings are different from those obtained with linear distractors and osteotomies.

The selection of the vector of distraction is also a critical decision. The corticotomy and the position of the pins determine the distraction vector. It is different in each patient depending on the grade of mandibular hypoplasia. In patients with Grade I the pins must be placed perpendicular to the corticotomy, obtaining an oblique vector that produces a larger bone elongation in the angle and a minor bone elongation in the alveolar ridge. In patients with Grade II-A hypoplasia the distraction process must remodel and elongate the angle and the initial portion of the ascending ramus (Fig. 25.3). For this reason the pins must be inserted in an intermediate position between a vertical and oblique distraction vector. This group of patients represents the vast majority of our clinical series.

In patients with Grade II-B hypoplasia the corticotomy is placed horizontally in the base of the ramus. The pins must follow a strict vertical distraction vector in order to obtain more elongation in the hypoplastic ascending ramus.

Our distraction protocol includes a latency period of 4–5 days. Distraction commences at a rate of 1.0 mm/day (distraction period). This period is continued 4–6 weeks until we achieve the desired mandibular lengthening with the correction of the facial asymmetry, chin position, level of the oral commissure, and the changes in the dental occlusion. A very important point is to produce an overcorrection in the group of growing patients. In the preoperative period the patients present a marked lateral deviation of the mandibular teeth to the affected side. After the distraction has been completed, the deviation of the mandibular interincisal line is reproduced in the

opposite direction in the contralateral side as a result of overcorrection. Also, a posterior open bite and a crossbite are produced immediately after distraction; for these reasons, posterior bite blocks, dynamic appliances and standard orthodontics maneuvers will easily control the occlusal changes and will produce some degree of dental occlusion stability. Bite blocks were required to maintain the posterior open bite; the blocks were gradually reduced to allow vertical descent of the maxillary dentoalveolus. We must always avoid the production of occlusal disasters as this situation will be impossible to correct later with standard orthodontics procedures.

The consolidation period is about 8 weeks or more. It depends on the age of the patient as well as the amount of bone lengthening. In an adult patient with a severe hypoplastic ramus, it can be prolonged for 16 weeks until there is radiographic evidence of mineralization. At this time the device is removed under sedation.

Age is also a critical factor in the treatment of a hemifacial microsomia patient with mandibular distraction. During the 1990s the international criteria

was to indicate the procedure at an age of 5–6 years, including the more severe cases. This decision was established according to the amount of mandibular bone stock and the feasibility of using intra- or extraoral distraction devices. Actually, we know that patients under 5 years of age, presenting a severe or moderate deformity, are not good candidates to elongate the mandible with intraoral devices. The procedure will result in a failure to achieve the ideal distraction vector, secondary occlusal disasters, and the need for future surgical interventions.

In our clinical series aesthetic results were excellent. The facial symmetry was restored with descent of the buccal commissure to the level of the contralateral normal side; the menton became horizontal and



Fig. 25.4. **a** Preoperative frontal view of a 9-year-old girl with a Pruzansky II-A hemifacial microsomia showing facial asymmetry and chin deviation to the affected side. **b** During the distraction process, 2 weeks after the initial 16 mm of mandibular elongation showing an early improvement of the facial asymmetry. **c** Six months after 24 mm of mandibular distraction showing facial symmetry with descent of the buccal commissure and medialization of the chin. **d** The patient 12 years later showing stability in the clinical result. Facial symmetry: a chin position has been stable in the long term result

was located at the midline (Fig. 25.4a–d). In all our patients we noted an increase in the soft tissue envelope bulk. This simultaneous soft tissues expansion is the most important factor in obtaining the correction of facial asymmetry and it is more observed during the consolidation period. At this time, the device serves as an external fixator and the muscles of mastication produce contraction forces over the regenerate bone allowing the molding of the callus with the increase of the bigonial distance by the muscles' pull.

Our clinical series also includes a group of patients with hypoplasia Grade II-B treated between 18 months to 3 years of age. Undoubtedly this group of children has better craniofacial growth and long-term clinical stability in the soft tissues. After overcorrection, the use of dynamic orthodontics appliances and bite blocks corrected the occlusal changes and the vertical dimension of the maxilla. All of these changes occurred very fast and almost spontaneously during a period of 2–3 months, compared with children treated over age of 6 years. For this reason every day, more and more, we are indicating the procedure at early ages. It is also important to mention that permanent dental injury or neurovascular damage did not occurred. Elongation was done by the parents with minimal dis-

comfort during the night without disturbing the children's sleep. These children represent the group of patients with the best results and better tolerance to the method.

Twelve percent of the patients with hypoplasia Grades II-A and II-B treated after 6 years of age required a second distraction procedure. The main reasons for a secondary procedure were: erroneous distraction vector, incomplete consolidation period, and a nonsatisfactory overcorrection criteria. These patients presented an early tendency to retake the original condition of facial asymmetry and malocclusion as it was observed after 2–3 years of follow-up (Fig. 25.5a–i).

If the patient is 12 years old or more at the time of the second bone elongation, we indicate a simultaneous maxillo-mandibular distraction. This technique is also indicated in the adult group of patients as a primary surgical procedure (Fig. 25.6a,b). Additional to the mandibular distraction, we include a subperiosteal dissection of the maxilla, then a complete horizontal osteotomy is made at the level of the pyriform area in both sides. The pterygomaxillary junction is freed with a curved chisel and Rowe forceps on the affected side only. Rowe forceps may be used softly to

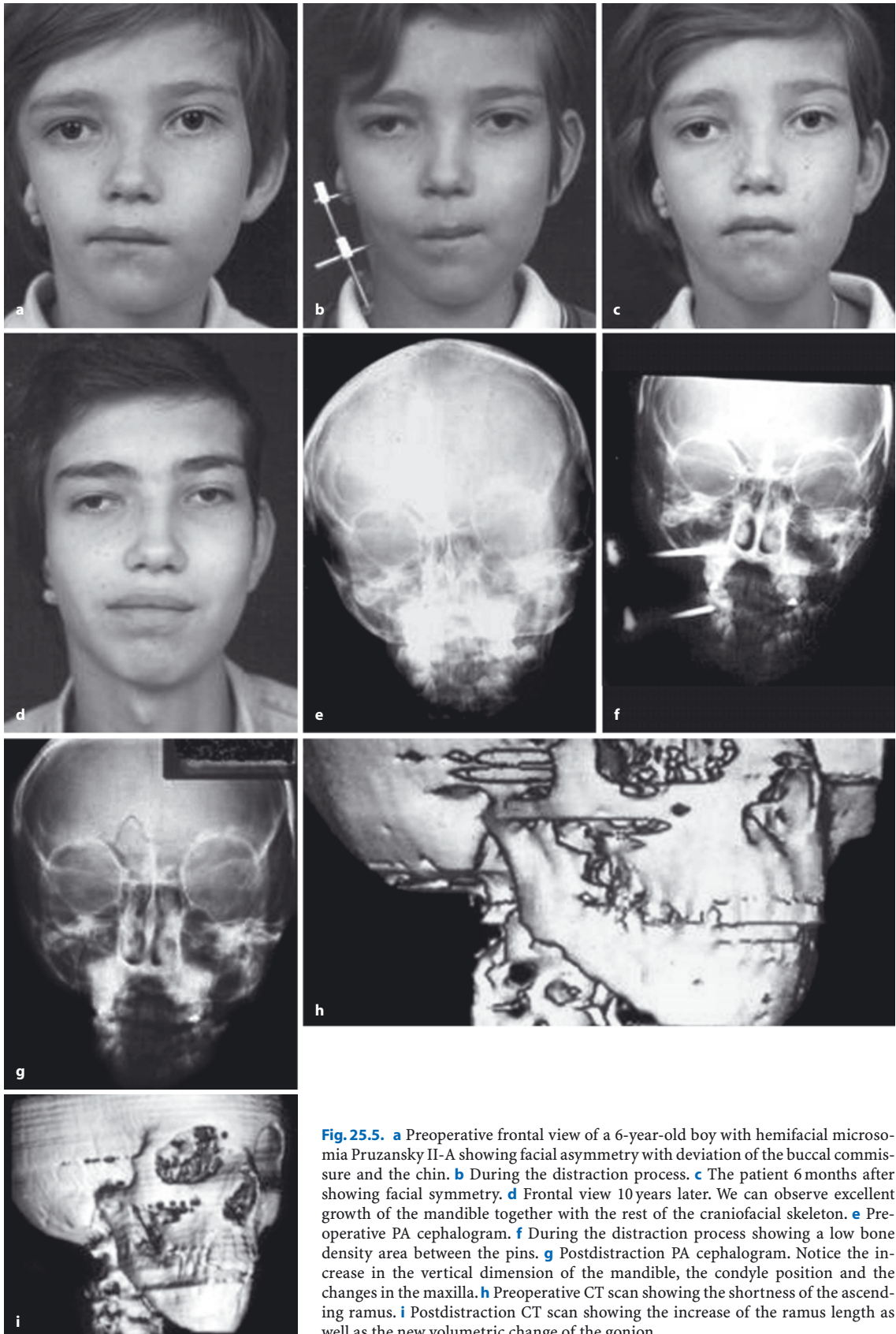


Fig. 25.5. **a** Preoperative frontal view of a 6-year-old boy with hemifacial microsomia Pruzansky II-A showing facial asymmetry with deviation of the buccal commissure and the chin. **b** During the distraction process. **c** The patient 6 months after showing facial symmetry. **d** Frontal view 10 years later. We can observe excellent growth of the mandible together with the rest of the craniofacial skeleton. **e** Preoperative PA cephalogram. **f** During the distraction process showing a low bone density area between the pins. **g** Postdistraction PA cephalogram. Notice the increase in the vertical dimension of the mandible, the condyle position and the changes in the maxilla. **h** Preoperative CT scan showing the shortness of the ascending ramus. **i** Postdistraction CT scan showing the increase of the ramus length as well as the new volumetric change of the gonion

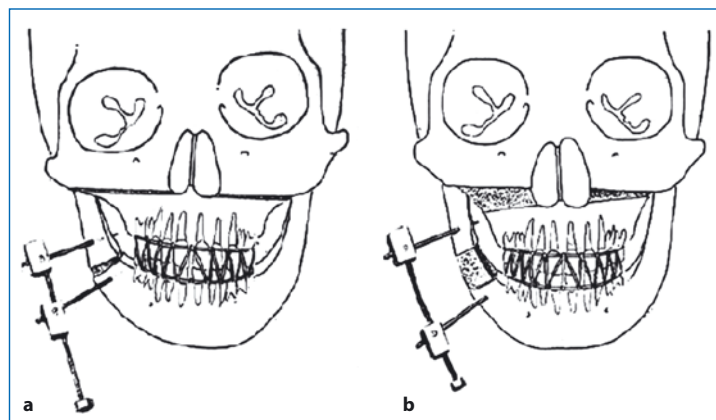


Fig. 25.6. **a** Diagram showing the mandibular corticotomy with the device in site. An additional horizontal osteotomy has been performed over the maxilla. After 5 days an intermaxillary fixation is indicated with the aim of obtaining a simultaneous elongation of both bone structures. **b** Areas of new bone formation over the mandible and the maxillary

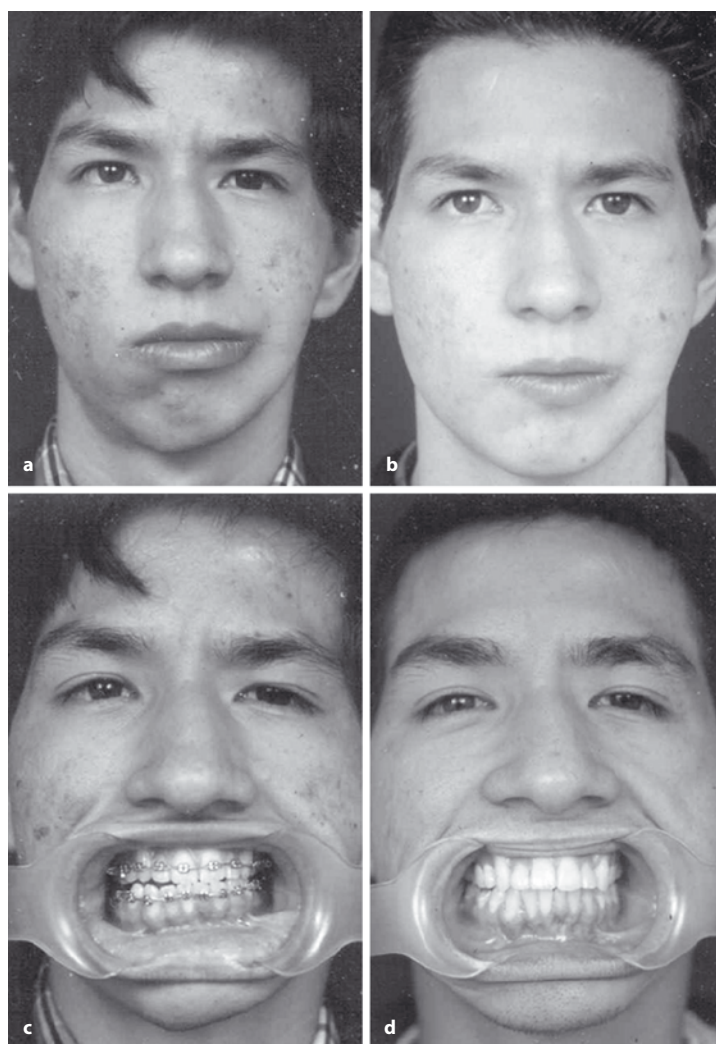
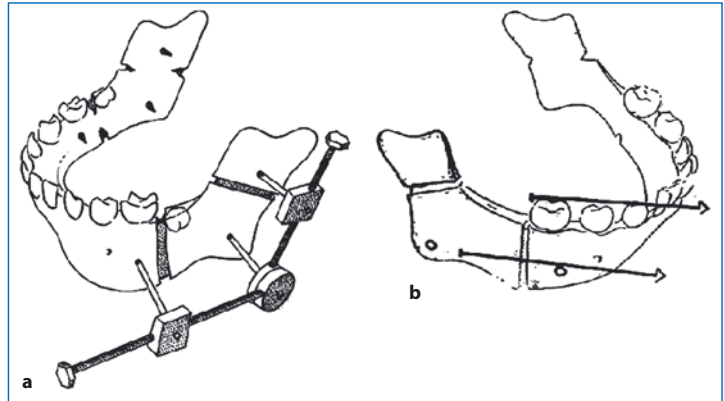


Fig. 25.7. **a** Preoperative frontal view of an adult patient with left hemifacial microsomia. **b** Six months postdistraction frontal view after a simultaneous maxillo-mandibular distraction. **c** The patient showing the operative occlusion. Notice the obliquity of the occlusal plane. **d** Six months after distraction the patients shows its occlusion with the correction of the occlusal cant

Fig. 25.8. **a** Diagram showing two corticotomies for bidirectional elongation. The central osteotomized bone fragment must be large enough to avoid fracture and back displacement. **b** The horizontal vector always has to be parallel to the occlusal plane in order to avoid the production of an anterior open bite or other occlusal disasters



assess the completeness of the osteotomy but no attempt is made to mobilize the midface. The pterygo-maxillary junction on the unaffected side remains intact and it will serve as a pivot to safely produce the midface elongation-rotation movements on the affected side.

The result shows that with the mandibular elongation, the maxilla follows the mandible changes, achieving simultaneous elongation, medial rotation, and advancement. Preoperatively the deviation of the occlusal plane from the horizontal varied from 12° to 18° and after the combined distraction procedure the clinical and orthodontic correction was obtained with a minimal slanting of 1–2° in the long-term follow-up.

Preoperatively, the vertical dimension of the maxilla was very short and the last mandibular molar was in a very close relationship with the malar bone. Postoperatively, the vertical maxillary increase from 16 to 15 mm, representing a 90% correction when compared with the unaffected side. All of this new bone formation results in a simultaneous correction of the oblique nasal floor, as well as the deviation of the nasal septum changing into the more normal position, increasing the volume of the nasal cavity. The pre-existing dental occlusion has been preserved; after the elongation, the superior and inferior midline incisors have changed and now are positioned in the central line of the face (Fig. 25.7 a–d).

The chin is always medialized by the distraction but in adults it is not always possible to achieve a normal central position; for this reason this procedure does not eliminate the need for a sliding genioplasty or another extrasurgical procedure, like a free dermis-fat graft, at the time of the removal of the device to improve the aesthetic final result.

25.1.2 Micrognathias

Patients with micrognathias present a different problem because of the bilateral deformity and because both the mandibular body and the ascending ramus are affected, requiring bidirectional and bilateral distraction. This concept is also true for patients with Pierre Robin, Nager, Treacher Collins, and bilateral hemifacial microsomia syndromes. In these cases two corticotomies were done: a vertically oriented one in the mandibular body and a horizontally oriented one in the ascending ramus. The pins used are as follows: a central one is introduced at the mandibular angle between the two corticotomies, a second into the mandibular body, and a third into the central aspect of the ascending ramus. One bidirectional device is used on each side, each one with two distraction plates to allow independent and more precise elongation of each segment, using the central pin as the fixed pivot for both of them (Fig. 25.8 a, b).

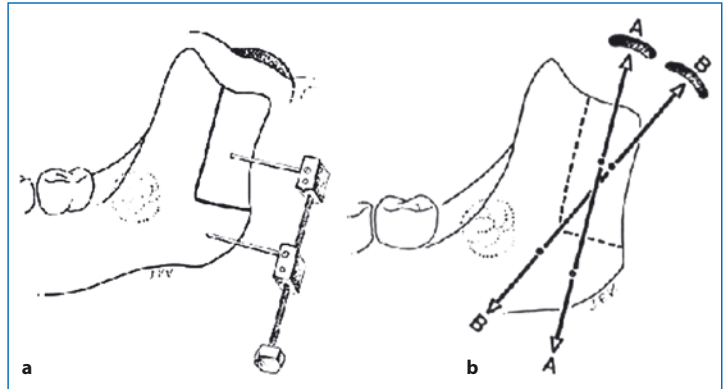
Most of these patients present the typical “bird face” deformity with deficient soft tissue at the lower third of the face and at the neck, absence of the neck angle, and shortened suprahyoid muscles. With bone distraction, all the tissues from skeleton to skin are simultaneously elongated without the inconvenience of osteotomies or skin expansion (Fig. 25.9 a–h). After conventional osteotomies and bone grafts, the muscles and the tight skin envelope present a limiting factor often requiring multiple procedures and seldom achieving optimal aesthetic results. Tissue expansion increases the size of the skin cover, but other soft tissues such as muscles, vessels, and nerves remain unchanged. The overall functional and aesthetic results with bidirectional mandibular distraction have been spectacular. The neck takes a normal shape with a well-defined angle. The muscles and soft tissues of the floor of the mouth together with the masticatory muscles, giving a better aspect of the neck. The chin takes a more prominent position.



Fig. 25.9. **a** Frontal view of a 22-year-old girl during the distraction process of a bilateral and bidirectional mandibular elongation. **b** Panorax view showing the four independent vertical and horizontal distraction vectors. In asymmetrical micrognathias one mandibular body can be elongated more than the contralateral in order to obtain symmetry. **c** Preoperative frontal view of the patient showing the asymmetrical micrognathia secondary to a condyle fracture during the early years of life. **d** Frontal view 3 years later of the mandibular distraction.

Facial symmetry has been restored. **e** Preoperative lateral view showing the classic “bird face.” **f** Lateral view 3 years later showing the new mandible with a nice elongation of the soft tissues over the superior third of the neck. **g** PA cephalogram 3 years later showing a very well-defined mandibular structure. **h** Lateral cephalogram 3 years later. After mandibular elongation the procedure has obtained a normal anatomy with an ascending ramus, gonion, mandibular body, and chin. The use of proper distraction vectors has achieved a normal occlusal relationship

Fig. 25.10. **a** Diagram showing the “L osteotomy” over the ascending ramus with the device in place. **b** The distraction vector most to be directed to the empty glenoid fossae



25.1.3 The Temporomandibular Joint Ankylosis

Studies of the effect of mandibular distraction on the temporomandibular joint have shown that the condyle assumes a more normal anatomic size, shape, and position after distraction [11]. In fact the process of distraction is beneficial to the condyle structure and its position in the glenoid fossae.

Regardless of the etiology of the joint ankylosis, it can be treated using the concept of bone transport and distraction to generate a neo-condyle and to obtain a normal mandibular anatomy. In extra-articular ankylosis through a vestibular incision and subperiosteal dissection, a L-shaped osteotomy is made from the medial aspect of the projected condyle down to the posterior aspect of the ramus. Two pins are inserted, one in the osteotomized bone fragment and a second at the angle. The distraction vector is directed to the empty glenoid fossae (Fig. 25.10a,b). After a 5-day latency period, the osteotomized segment of bone is distracted and transported in a vertical direction until achieving the planned condyle lengthening. Usually 14–20 mm is enough to achieve a similar dimension with the contralateral side. Immediately the patients begin with open-close exercises and during this activation period, the edge of the neo-condyle is remodeled into a smooth, rounded surface. Fibrocartilaginous tissue at the leading edge of this segment acts as a pseudodisc.

If the patients present an intra-articular ankylosis (true bony fusion), an arthroplasty is performed through a preauricular incision. The surgeon must release the bony fusion and he will avoid bone resection reducing the ramus height. The abnormal bone is remodeled with a burr and once this maneuver is completed, the opening of the mouth is accomplished with the assistance of a special forceps. Then a very fine Silastic layer (less 1.0 mm) is introduced as a cap on the top of the projected condyle and fixed to the sur-

rounding scar tissue with two resolvable stitches. We close the incision and then we change into the intra-oral route for the L-shape osteotomy.

This method produces a vertically elongated mandible with a functioning nonankylosing temporomandibular joint in the short term. The interincisal distance obtained ranged from 35 to 45 mm in the adult group. This technique will probably become the treatment of choice of temporomandibular joint ankylosis. Better diagnostic methods and understanding of etiology will be necessary in the near future to determine the efficacy of this process.

25.1.4 Cleft Lip and Palate Patients

The etiology of midface deficiency in cleft patients is uncertain. The absence of Class III malocclusion in untreated clefts suggests that previous cheiloplasty and palate repair may be related to this condition [34]. Ross [35] in a longitudinal survey of 528 repaired cleft patients from 15 centers around the world, demonstrated that approximately 25% develop maxillary hypoplasia that didn't respond to orthodontic procedures alone.

At the time of treatment, the ages of patients ranged between 6 and 12 years (mean 8 years), and all of them demonstrated clinical and radiographic evidence of moderate to severe underdevelopment of the maxilla (sagittal, vertical, and transverse). They also presented with a flattened middle third of the face and Class III malocclusion, with partial or total anterior and posterior crossbite.

The severity of maxillary hypoplasia ranged from 2 to 10 mm of A-P discrepancy. Class III malocclusion with anterior and posterior crossbite, deep overbite, and in some patients, vertical skeletal open bite was observed.

Preoperative orthodontics are mandatory to obtain a minimal occlusal stability. The following orthodon-

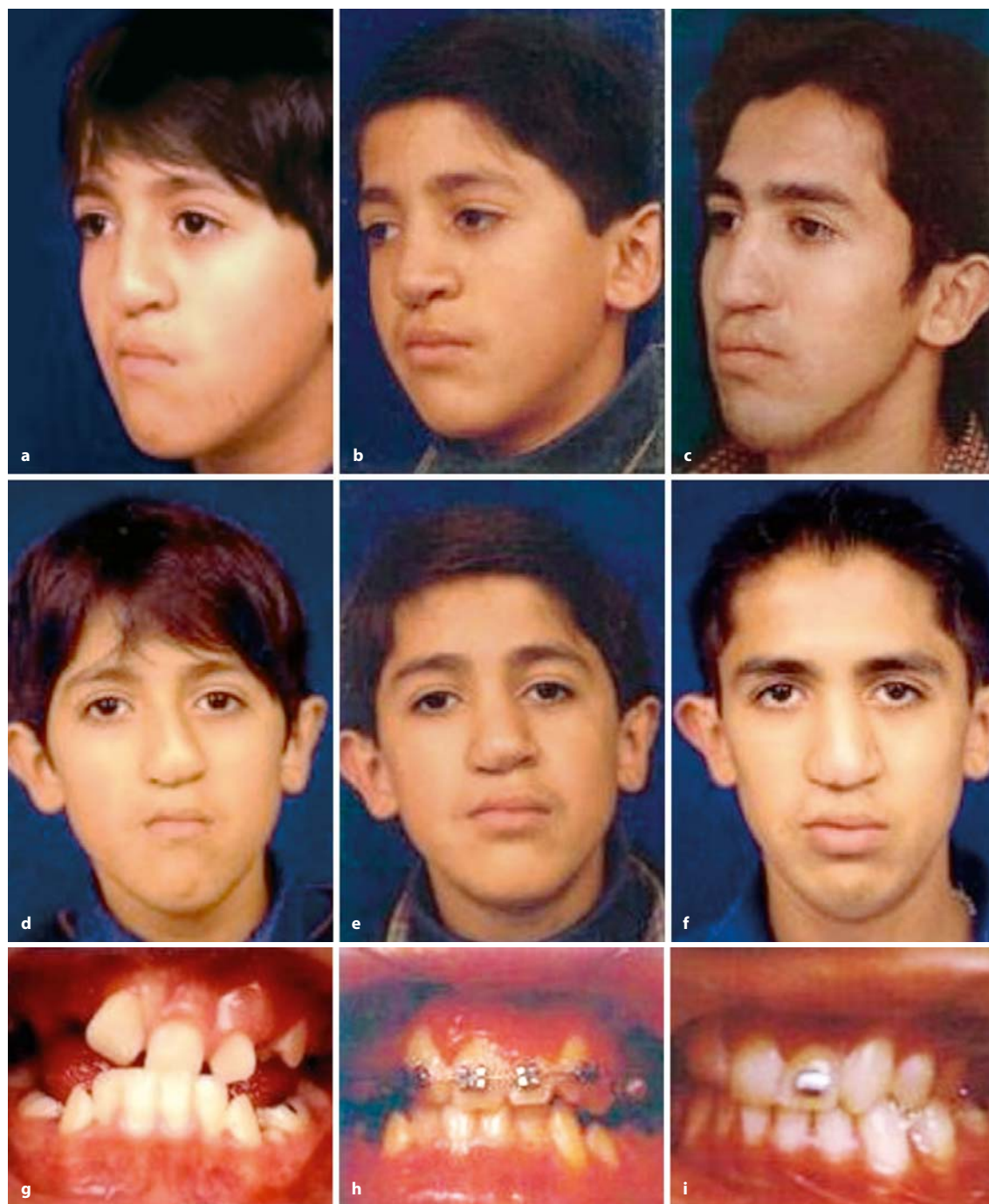


Fig. 25.11. **a** Preoperative three-quarters view of a 9-year-old boy with unilateral cleft of lip and palate and moderate midface retrusion. **b** Postoperative three-quarters view 1 year before 6 mm. of advancement and 3 mm of vertical elongation with maxillary distraction. Notice that the new position of the maxilla produces a minor anterior mandibular rotation. The middle third of the face has been elongated, producing a more appropriate balance of the middle and inferior thirds of the face. **c** A postoperative three-quarters view 9 years later. **d** A preoperative frontal view. Because the middle third of the face is retrud-

ed and short, an exaggerated anterior rotation is produced in the mandible. The patient looks prognathic and with a very long inferior third of the face. **e** The face in its thirds looks well proportioned and balanced. **f** Clinical stability with nice aesthetic results in the long term. **g** Preoperative occlusion showing negative overjet and posterior crossbite. **h** Orthodontic treatment continues that includes the alignment of the permanent incisors in the cleft area and conventional orthodontics. **i** The final occlusion

tic requirements were fulfilled prior to maxillary distraction: skeletal and/or dentoalveolar transverse expansion of the maxilla, alignment of permanent incisors in the cleft area in some patients and bone grafting at 8–10 years old in other cases [36–38].

Prior to surgery a modified quad-helix fixed appliance was placed in the palate. This is made of a simple lingual arch attached to bands placed on the second deciduous molars. Anteriorly the arch maintains passive contact with the incisors through an acrylic button. In order to maintain maxillary expansion during the advancement, an extra transverse arch is fixed to both premolar bands which in turn provide the support for the vestibular hooks.

Under general anesthesia two maxillary vestibular incisions 3–4 cm long are made, leaving a 2-cm central mucosal bridge between the incisions. Subperiosteal dissection is performed from the pyriform fossae to the lateral maxillary buttress on each side exposing the anterior and lateral aspects of the maxilla to the level of the infraorbital nerve and above the canine roots. Dissection on the nasal floor is limited to the lateral part only. The nasal septum and the maxillary cleft gap remain intact. Using a side-cutting burr, a horizontal osteotomy is performed above the roots of the canines and molar tooth buds as identified on the posteroanterior cephalogram and panorex.

Bilaterally, using a 7-mm chisel, the osteotomy is extended into the maxillary buttress, reaching the pterygomaxillary region. Dysjunction with Rowe forceps is not performed, thereby avoiding unnecessary bleeding and uncontrolled fractures in the pterygomaxillary junction. At this time the surgeon must assess maxillary mobility by applying gentle downward pressure to the LeFort I segment. The mucosa is then approximated and closed with absorbable sutures. Following this minimally invasive technique, the hypoplastic maxilla is ready to be mobilized. As the nerve function and vascularity have been preserved, the mechanical distraction forces will stimulate the functional matrix to produce new bone formation.

Using the fixed intraoral appliance system as an anchorage for a modified “Petit” facial mask supported by the forehead and chin, distraction forces are initiated on postoperative day 5. Two intraoral elastic bands on each side are linked from vestibular hooks to a bar on the mask. No other fixation is required. The initial mechanical force used for distraction is about 900 gm. The facial mask is used mainly at night and at some periods during the day for a total of 16–18 h every day.

When the vertical length of the maxilla is deficient, a combination of forward and downward distraction forces can be used to simultaneously advance and lengthen the maxilla.

Advancement of 2–3 mm is achieved each week. During the second and third week of distraction, the presence of palatal scars may produce difficulties in further advancement. At this stage, it is critical to maintain the same elastic force in order to weaken the resistance of the fibrous tissue. If necessary, the elastic bands, which are initially parallel, may be crossed in order to increase the force of A-P distraction. In this manner, the projected maxillary advancement and a satisfactory Class II molar relationship is obtained. At this point, the amount of force is decreased to one elastic band on each side (450 gm) for another 2 months (consolidation period), in order to maintain the maxilla in its new position.

Radiographic evidence of new bone formation is observed at approximately 8–10 weeks postdistraction. The bone step formed by the horizontal distraction is replaced with mature cortical bone. There is also evidence of a secondary area of new bone formation at the pterygomaxillary junction. These findings attest to the skeletal stability and will diminish the risk of relapse.

The projected advancement was achieved in all of the patients with results varying from 4 to 12 mm (mean 7 mm). The aesthetic results were excellent (Fig. 25.11 a–f). The soft tissue profile changed from concave into a well-proportioned orthognathic face. Changes also included an increased nasolabial angle, a more anterior projection of the upper lip, and an improved lip relationship.

Dental occlusion was changed to a distal sagittal molar relationship with moderate overjet (Fig. 25.11 g–i) with significant downward clockwise rotation of the mandible. Cephalometrically, a counterclockwise rotation of 2°–7° was achieved. The increase in the ANB angle varied 1.4°–7.4°. After distraction was completed, it was essential to stabilize the maxilla with a fixed palatal arch, in order to proceed with the orthodontic treatment such as alignment of permanent incisors in the cleft area and to correct the minor dental irregularities in the cleft site.

25.1.5 Craniosynostosis

In this clinical series different osteotomies are included: Le Fort III, fronto-orbital advancement and monoblock. The osteotomies were performed using a subcranial or an intracranial approach. The intracranial route is always assisted endoscopically in order to perform a clean dissection of crista galli and cribriform plate, to assess for hemostasis and to verify dura mater disruptions, and most importantly to preserve vascularity between the dura and the skull. The surgical technique uses several basic procedures of the classic craniofacial osteotomies. Under general endo-

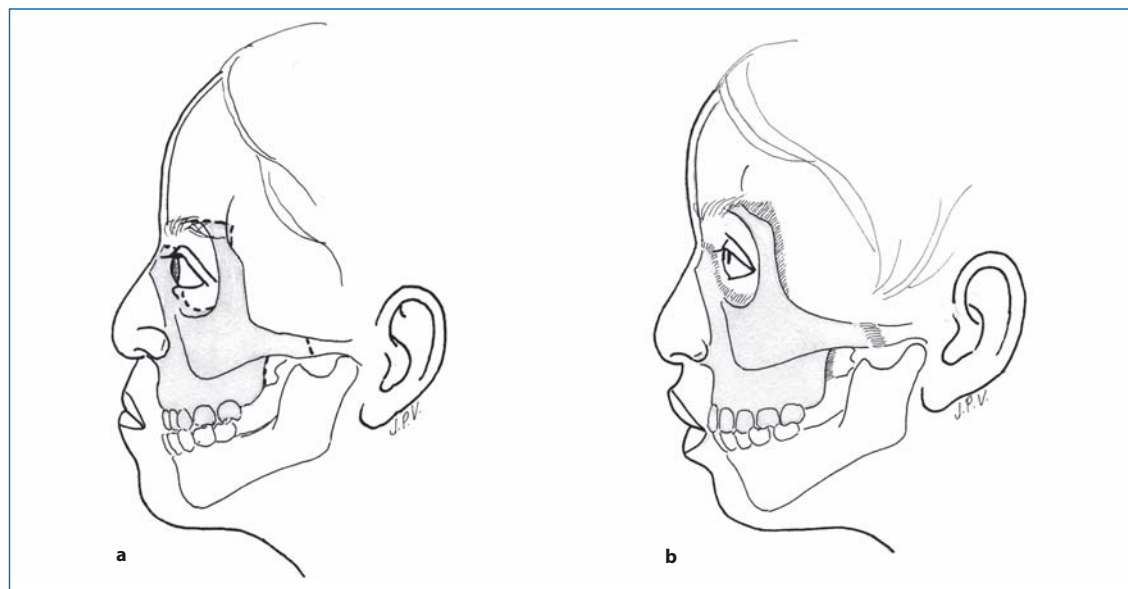


Fig. 25.12. **a** Osteotomy line of the modify LeFort III Osteotomy. At the nasal bones and at the supraorbital border the osteotomy preserves some millimeters of bone attachments, avoiding the production of long noses and bone steps at the fronto-orbital region after the advancement. Malocclusion in Class III is observed. **b** After advancement, new bone formation is ob-

served along the osteotomy lines, most importantly in the lateral aspect of facial framework, the lateral orbital wall, the zygoma, and pterygomaxillary area. At the nasal bone and the superior orbital edge, new bone formation is minimal when compared with the rest of the osteotomies. The occlusion has been corrected

tracheal anesthesia, exposure of the midface and the orbits is obtained through a standard coronal incision.

The dissection is carried over the periosteum, and at the temporal level the dissection extends to the deep temporal fascia so that the soft tissue envelope can be reflected off the lateral orbital rim and zygoma. At this time the temporal muscle is sectioned below the temporal crest and with a limited dissection 15 mm wide, we reach the lateral orbital wall until the retromalar area. In fact we preserve the vast majority of the temporal muscle insertion.

Then the dissection is extended to the orbit superiorly at the orbital roof 8–10 mm from its apex, laterally until the inferior orbital fissure is clearly visualized, and medially until the nasolacrimal groove without disruption of the medial canthal ligament.

In a patient with previous fronto-orbital advancement and cranial vault surgery with midface hypoplasia and associated Class III malocclusion (Fig. 25.12 a,b), a subcranial Le Fort III is indicated. The osteotomy is designed including the lateral half of the supraorbital rim. Here the osteotomy has to preserve 6–8 mm of bony union at the medial portion. Then the osteotomy is followed to the external aspect of the lateral orbital wall and extended down until reaching the junction of the maxilla and the pterygoid plate. At this point an interpterygomaxillary disjunction is

done with a 12-mm chisel. The intraoral classic route is avoided. Then an oblique osteotomy is performed at the zygomatic arch.

Following intraorbitally the bone cut at the infero-lateral angle, it extends from the inferior orbital fissure along the orbital floor reaching medially the posterior aspect of the nasolacrimal groove. This maneuver allows the advancement of the nasolacrimal apparatus and medial canthal tendons anteriorly with the midface.

Over the nasal base one horizontal osteotomy is performed connecting the medial orbital wall osteotomies in both sides. Craniofacial disjunction is accomplished with Rowe Kiley forceps. The surgeon must avoid fracturing the remaining 6- to 8-mm bone union at the supraorbital rim. This incomplete osteotomy will act as a pivot and will avoid bone steps in the fronto-orbital area after the advancement.

The distraction devices are placed in the midface in two different ways: simply supported behind malar bone or anchored with a hook to the malar-orbital edge. The device is a submerged type and is made of one hollow plate with a central perforation to allow the entrance of a screw running free, 25–30 mm in length. After the screw thread the rest of the device has a smooth surface and can be easily positioned through the temporalis muscle reaching the malar's

Fig. 25.13. **a** A 4-year-old girl with Crouzon disease. A fronto-orbital advancement has been performed at age of 12 months. Exorbitism and severe midface retrusion with sleep apnea are still unresolved problems in this patient. **b** At the end of the distraction process. Malar-zygoma advancement was 19 mm, solving the exorbitism. The maxillary position has corrected the airway problem. Facial proportions now are well balanced. **c** Preoperative lateral cephalogram showing the retruded midface and the rigid fixation system, mini-plates and wires used in the fronto-orbital area during the surgical procedure. Notice the fine airway secondary to the severe maxillary hypoplasia. **d** Post-distraction lateral cephalogram showing the bone advancement and the position of the devices. An oblique vector has produced the bone advancement. It has been gradual and different at the orbital level and at the maxilla level



back or anchoring to the bone edge with an extra hook. The hollow plate is screwed to the parietal bone.

To advance the midface, a horizontal vector is used. The vector has to be parallel to the occlusal plane and the midface is advanced until a Class II molar relationship is obtained, and/or, the zygoma-malar-orbit advancement corrects the exorbitism.

If the patient's midface is short vertically, then an oblique vector is the logical indication. Simultaneously, the bone movement will correct the midface retrusion and will lengthen its vertical dimension.

The obtained subcranial advancement ranges from 18 to 30 mm. This distance has been measured at the orbito-malar region. It is very important to observe that the maxillary advancement ranged from 7 to 12 mm and at the supraorbital region varied from 10 to 16 mm. Certainly we have obtained a gradual bone advancement that is different at the orbit, at the malar, and at the maxilla allowing the correction of the exorbitism and of the midface hypoplasia.

This gradual advancement is generated by an additional rotational movement in the lateral aspect of the facial framework, more accentuated at the zygoma-malar area. Probably this additional movement offers a more satisfactory correction of the exorbitism and avoids the production of the long "French noses" as typically are obtained after Le Fort III advancements.

Aesthetic results were excellent and a final facial appearance is comparable with normal, good-looking individuals (Fig. 25.13 a–d).

In a primary syndromic craniosynostosis patient (Apert, Crouzon, Pfeiffer, etc.) presenting with severe midface hypoplasia and secondary respiratory problems, the monoblock osteotomy is the indicated. The design of this osteotomy represents a "true monoblock" because frontal bone, both orbits, malars, and maxilla move anteriorly together in one single piece (Fig. 25.14a, b).

In these cases, the osteotomy is outlined at the frontal bone and the lateral aspect of the osteotomy runs down until it reaches the lateral orbital wall. It then follows in a manner similar to the subcranial route.

Two or three craniotomies are performed. Below the outlined osteotomy design, intracranial tunnels are dissected, introducing large fine gauzes in order to protect the meninges. Special attention must be paid at the tunnels at the supraorbital area as well as at the crista galli.

At this point a flexible endoscope is introduced into the tunnels and assessment for hemostasis is performed. Also, a safe intracranial dissection at the more dangerous area must be completed under vision; this maneuver is specially important in patients in which

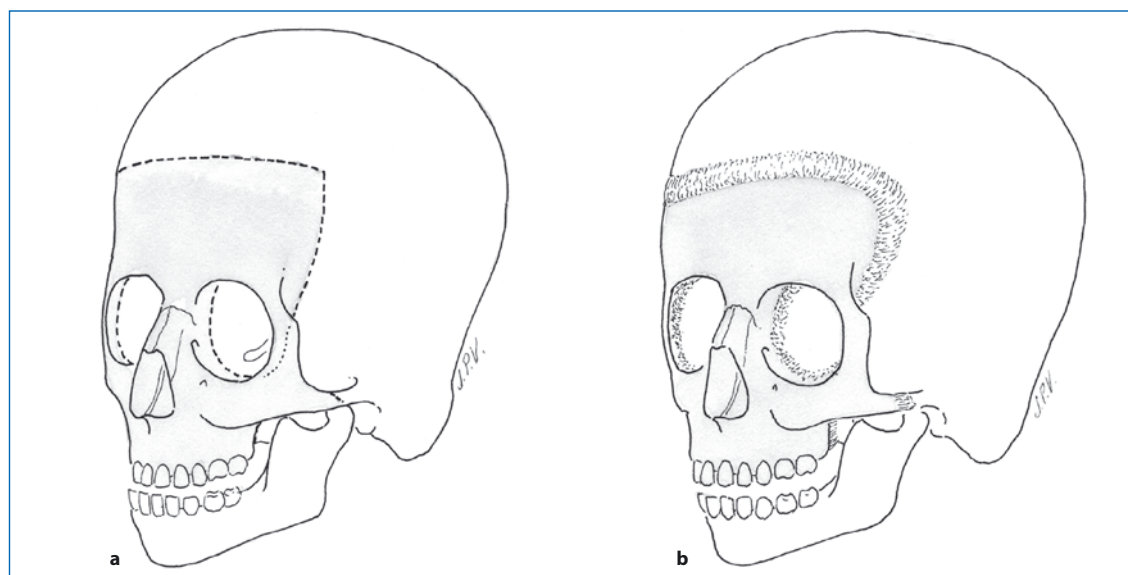


Fig. 25.14. **a** The “true monoblock” osteotomy. Doted line shows the single-piece osteotomy that includes the frontal bone, the orbits with their 4 walls, the lateral and medial walls,

and the floor, the pterygomaxillary junction, and the zygoma. **b** After distraction, new bone formation is observed along the osteotomies lines, included at the pterygomaxillary tuberosity

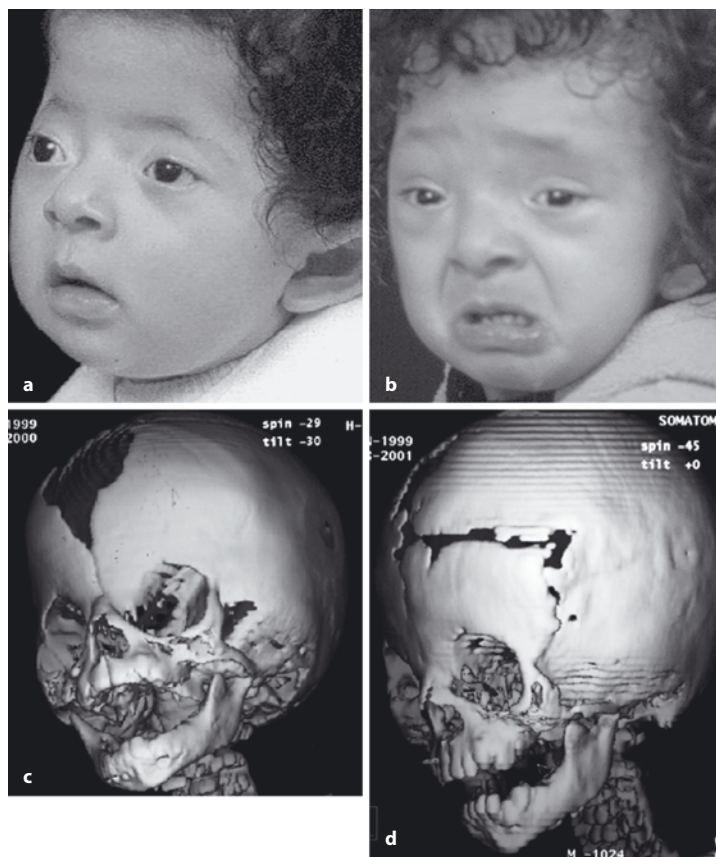


Fig. 25.15 a–d. **a** Pre-operative third quarter view of a 14 months old girl with Apert Syndrome showing a severe supra-orbital depression, fronto-orbital deformity and midface retrusion. **b** Post distraction clinical changes. The “true monoblock” has corrected frontal deformity, exorbitism and midface retrusion. Oxygen saturation is normal during sleep. **c** A preoperative CT scan of a 14 months young girl with Apert Syndrome, brachycephaly with a severe flattened supra-orbital region is observed together with severe midface retrusion. Oxygen saturation drops during the sleep of the patient. **d** Post distraction CT scan shows the new position of the mid-face with the simultaneous correction of the orbits position and the frontal bone after a “true monoblock” advancement. New bone formation is observed in the lateral aspect of the frontal bone, in the lateral orbital wall, the zygoma and at the pterygomaxillary area. On the frontal bone an extra remodeling is observed in the patient during the consolidation period. The brain expansion force produce a round shape on the frontal bone achieving a structure very closely to the normality

the dura is firmly attached to the internal cortex. The maneuver will avoid meningeal disruptions.

This sort of intracranial dissection preserves vascular attachments between the frontal bone and the dura along with the preservation of the bone vascularity. In reality we are generating bone flaps that will produce a healthy new bone formation along the osteotomy lines very quickly. The preserved vascularity will avoid intracranial dead spaces and secondary bone necrosis, often observed after the classic advancements with osteotomies.

The frontal osteotomy is performed with an oscillatory saw. A fine brain retractor is introduced into the tunnels over the gauzes and then the cut is completed safely. Over the orbital roof and the medial wall a curve osteotome is used to complete the osteotomy. Of significant importance is completion of the bone cut at the crista galli. This maneuver is done with two 7-mm chisels. The first one is introduced through the central craniotomy and endoscopically positioned in front of the crista galli, then with gentle hammering the osteotomy is begun. Immediately thereafter the second chisel is positioned exactly in front of the first one and the osteotomy is completed medially and laterally until the bone cut reaches the orbital roof.

At this point a Rowe Kiley forceps is used to complete the monoblock disjunction and maxillary disimpaction forceps can be used in an effort to facilitate the anterior midface advancement with simultaneous stretching of the overlying soft tissue envelope.

A pair of distraction devices are used to advance the whole bone mass in one piece. Using the extra hooks, the devices are anchored to the fronto-orbital union edge, through the temporalis muscle, and screwed to the parietal bone.

The postoperative course in this group of patients has been very satisfactory. Monobloc advancements ranged from 20 to 30 mm in the fronto-orbital region and from 10 to 18 mm in the maxillo-malar area. The exorbitism has been corrected in all cases. It is our impression that simultaneous advancement of the roof, the floor, and the medial and lateral walls of the orbit produces a more anatomical correction (Fig. 25.15a–d). In the frontal region, gradual advancement of the frontal bone has produced good results, both in the contour of the forehead and cranium and in the relationship with the facial skeleton.

Extradural dead spaces between the frontal bone and the frontal lobes has not been observed in any of our patients. Also, irregular contour in the forehead secondary to partial bone absorption has been eliminated with the distraction advancement.

We link together the orbito-cranial and maxillary problems because their primary malformation occurs simultaneously and its correction is critical to obtain excellent functional results.

25.2 Discussion

Distraction osteogenesis is a technique that is becoming popular for the reconstruction of deficient mandibles and has simplified the treatment for congenital mandibular hypoplasia. Technically, it is a minor surgical procedure that preserves the integrity of the nerves and vascular supply. Careful planning of the corticotomy and the position of the pins produces a distraction vector that follows closely the direction of normal mandibular growth [10–16].

Vertical and sagittal mandibular elongation was obtained in all patients. During the early stages of our clinical work, we assumed that the elongation would always result in a posterior open bite. However, it did not occur in many of the cases due to dento-alveolar adaptation at the maxillary level [17–21]. This vertical growth change was documented cephalometrically, suggesting that the maxilla liberated from the constricting effect of the small mandible and deficient soft tissue can reach its normal growth potential. Murray and Mulliken [1] have reported the same increased vertical maxillary growth after chondrocostal grafting of the ascending ramus.

An enormously important benefit of bone distraction is the simultaneous expansion of the surrounding soft tissue of the face [19, 20]. In fact, we were surprised to see the rapid descent of the buccal commissure to a normal position, the horizontalization of the chin, the increase in the distance between buccal commissure and the external canthus and in the distance between buccal commissure and the inferior orbital rim in the unilateral cases. Also, once the distraction is completed, we have observed some additional increase in the volume of the cheek, which is probably related to improved muscular activity.

All the patients with micrognathia presented the typical “bird-like” facial deformity with deficient soft tissue in the lower third of the face and the neck, causing absence of the neck angle, with shortened suprahyoid muscles. The overall aesthetic results obtained have been spectacular, beyond the both the patients’ and our expectations. The neck takes on a normal shape with a well-defined angle; the muscle and soft tissue of the floor of the mouth expand; the masseter insertion and the muscle volume increase, and the chin becomes more prominent.

Slow distraction of the mandible following corticotomies results in increased mandibular mass and length simultaneously with soft tissue (skin, muscles, ligaments, blood vessels, and nerves) expansion. This adaptation occurs over a period of 8–12 weeks. Elongation of the mandible results in a increase in size as well as changes in its positions and shape that would not be possible using conventional orthognathic surgery such as a sagittal split osteotomy procedure [39–41].

The early treatment of midface retrusion in children with C.L.P. has to be considered a major goal. Early correction of the maxillary hypoplasia minimizes the psychological problems as well as provides benefits related to improved occlusion, masticatory, and respiratory functions. Advancing the maxilla with distraction forces requires only a minimal surgical procedure preserving vascularity and nerve integrity. This technique presents exciting treatment possibilities eliminating the need for rigid fixation systems, blood transfusions, prolonged orthodontic treatment and intermaxillary fixation.

When the maxilla is properly positioned, it can grow in a near normal manner, allowing the tongue to assume a physiological position. In this situation, the permanent maxillary incisors erupt spontaneously into a normal overjet and overbite [36–38].

The use of distraction vectors are fundamental to achieve optimal advancements. Vectors are obtained through the combination of the position of the bar on the facial mask and the direction of the rubber bands. There are three possibilities of maxillary distraction vectors: upward, forward, and downward. The most commonly used is the forward one, in which the horizontal vector must be parallel to the occlusal plane to avoid producing an anterior open bite.

Overcorrection with a Class II molar relationship is always performed, especially in patients with severe deformity or in patients treated during 6–9 years of age. This point is critical because distraction does not alter the inherent cellular growth pattern. Following early maxillary distraction in patients with severe midface retrusion, the individual growth pattern often reverts to the original tendency of reduced maxillary growth and an anterior crossbite may become re-established. Overcorrection is therefore necessary to achieve an equilibrium between maxillary and mandibular growth. The use of intraoral functional correctors (Frankel III) also play an integral role by adding an additional stimulus for maxillary growth.

Gradual maxillary advancement with distraction produced functional changes affecting respiration. Nasal breathing was improved as well as the air flow and patency of the nasal airway. These changes are produced by an opening in the nasolabial angle and the increase of volume of the nasal and pharyngeal airway as demonstrated in the preoperative and postoperative lateral cephalograms.

Two relevant points must be considered to predict the difficulties with the midfacial distraction technique: first, the quality of palatal tissue and palatal scarring. Due to the tendency for retraction and relapse, fixed retention is mandatory. Secondly, preoperative skeletal and dentoalveolar transverse maxillary expansion, as well as dental compensations may be necessary to prevent occlusal instability. Clinical ex-

perience has proven that these factors determine the long-term stability of the maxillary advancement in this series.

Also, with maxillary distraction, a simultaneous soft tissue expansion is observed, and the skin, fat, and muscles have a more favorable peri-oral, peri-nasal and infraorbital distribution. The nasal base remains unchanged and the position of the lips, teeth, and tongue are in better relationship when the patient smiles.

In treating craniosynostosis, conventional osteotomy surgery is limited by the resistance of the soft tissue envelope which can be managed only by gradual advancement or multiple surgical procedures. The results of midface osteotomy advancement with rigid fixation typically fall short of the desired goal [42–48].

Is well known that the growth of the cranial vault is directly influenced by the brain, the growth of the orbit directly induced by the growth of the eye, and the growth of the maxilla linked to the eruption of teeth [49]. Normally, until 4 years of age the cranial base, like the vault, follows the rapid and even pseudotumoral growth of the brain. In addition, between the ages of 2 and 7 years the cranial base follows the development of the face; the later depends on dental eruption and masticatory movements [44].

These are very important observations to indicate early midface surgery in patients with craniosynostosis. Moreover, after 7 years of age the development of the frontal bone, the ethmoid, and the sphenoid depends on their pneumatization. The growth of the anterior cranial base appears, therefore, to be induced mainly by that of the brain and the maxilla.

These two last facts are very interesting to analyze since distraction osteogenesis in these patients produces areas of new bone formation over the anterior cranial base and the brain expansion after the procedure has produced a secondary remodeling of the frontal bone. The brain expansion molds the regenerate.

At present, when the operative risk has been reduced to the minimum, when most of the major complications can be prevented and minor complications can be controlled, and when good functional results are usually obtained, more of our efforts must be directed toward better cosmetic results. The ideal goal in craniofacial surgery would be to produce a normal appearance in all patients [27, 28, 30, 31].

Conventional osteotomy surgery is limited by the resistance of the soft tissue envelope, which can be managed only by gradual advancement or multiple surgical procedures [50–52].

The advantage of distraction in midface advancement is thus an increase in the amount of correction that can be achieved in a single procedure, enabling a more complete anatomic correction and possibly

sparing the patient multiple-stage procedures [28, 30–32].

Distraction osteogenesis also can offer excellent aesthetic possibilities. During the process the surgeon can perform some extra changes or adjustments during the elongation period, to finally obtain the proper position of the different segments. In fact, we can obtain a harmonious relationship between the thirds of the face, producing a normal appearance in many patients.

Other advantages to treating a craniosynostosis with distraction are: it allowed the advancement to be performed without the need of bone grafting, thereby decreasing the length of the procedure and the donor-site morbidity. It had significantly lower relapse rates than standard midface advancement in cleft lip and palate [24–26], presumably because the soft tissue, which traditionally resists midface advancement, was also gradually distracted along with the bone. The lack of initial mobilization significantly decreased the time needed for postoperative recovery. There is a significant decrease in blood loss and postoperative pain after this procedure, and patients are typically able to be discharged from the hospital earlier. Gradual advancement using distraction allows the displacement of skeletal fragments over greater distances because the soft tissue envelope is allowed to accommodate gradually. The magnitude of advancement is almost twice that obtained with traditional advancement procedures and has the possibility of adding a vertical midface lengthening, correcting the shortness of this structure.

All of these features will diminish morbidity and will produce excellent functional changes and the possibility of better craniofacial growth.

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26 Cleft-Orthognathic Surgery

JEFFREY C. POSNICK, PAUL S. TIWANA

26.1 Introduction

The child born with cleft lip and palate requires individualized care within the context of a treatment plan that includes a thoughtful staged reconstruction. Cleft reconstruction should be carried out effectively to prevent exhaustion of the family's energy and resources and to limit secondary iatrogenic deformities [1–10]. When the clefted upper jaw grows poorly, the primary skeletal deformity is characterized by a flat/retrusive appearance of the midface and Angle Class III malocclusion (Figs. 26.1–26.4). Our goal for all children born with a cleft lip and palate is for them to reach adolescence with normal function and without negative attention being drawn to their original malformation [11–21].

26.2 Integrated Team Approach

In addition to performing soft tissue and skeletal procedures, the *cleft surgeon* plays a primary role in coordinating the patient's care from infancy through adolescence and beyond. The *orthodontist* provides interceptive treatment in childhood, identifies abnormal growth patterns of the facial skeleton, and carries out final orthodontic treatment in conjunction with orthognathic surgery when required [22–25]. The *speech pathologist* plays a critical role in speech assessment, which may require a nasoendoscopic-guided examination [26]. Before orthognathic surgery, a speech evaluation is necessary to characterize both velopharyngeal function and articulation errors [26, 27]. Velopharyngeal closure that is adequate before surgery may become borderline, and velopharyngeal closure that is borderline may become inadequate after surgery [22]. Surgical and orthodontic management of crossbites, open bite, cleft-dental gaps, and residual oronasal fistulas will predictably relieve sibilant articulation distortions [27, 28]. The *general den-*

tist and the other dental specialties provide comprehensive preventative dental care, and input regarding restorative needs. The *otolaryngologist* provides treatment in the early years for middle ear infections. During adolescence, identification and then surgical correction of septal and inferior turbinate pathology is important to relieve obstructive breathing [29–31].

26.3 Timing

Correction of the cleft jaw deformity is planned for when the permanent dentition is fully erupted, the arches have been orthodontically leveled and aligned, and maxillomandibular growth is complete [12–15, 32, 33]. Final transverse, horizontal, and vertical growth of the maxilla and mandible cease at varied chronological ages. Most cleft patients are “normal” mandibular growers. In general there is no need to wait until late into adolescence to complete definitive jaw surgery. The use of epiphyseal plate closure documented on hand wrist radiographs, or cessation of maxillofacial growth documented on sequential cephalometric radiographs taken at 6-month intervals can be helpful to determine skeletal maturity. In the past, some surgeons have proceeded with mixed dentition cleft–Le Fort I advancement procedures. Wolford [4] has shown that if *very early* jaw surgery is undertaken in a cleft patient, revision orthognathic surgery will often be required once skeletal maturity is reached. Other risks of *very early* jaw surgery include injury to the developing permanent teeth, and the introduction of fibrous tissue and callus at the osteotomy site. If there are significant psychosocial or functional considerations, the benefits of “early” surgery may outweigh the risks (Fig. 26.2). The hoped-for advantage that with the use of the “DO (distraction osteogenesis) Technique” in conjunction with a Le Fort I osteotomy the maxilla would “grow” has not proven to be the case.



Fig. 26.1 a–i.

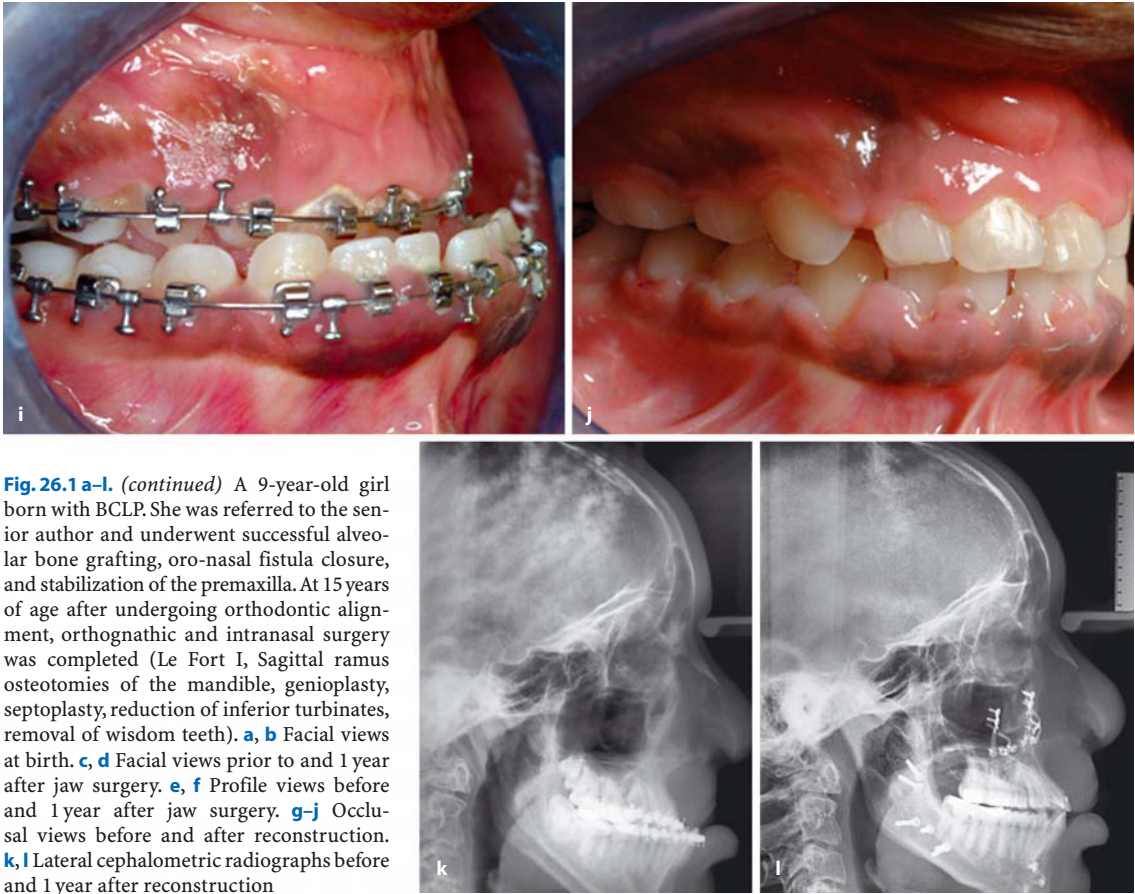


Fig. 26.1 a–l. (continued) A 9-year-old girl born with BCLP. She was referred to the senior author and underwent successful alveolar bone grafting, oro-nasal fistula closure, and stabilization of the premaxilla. At 15 years of age after undergoing orthodontic alignment, orthognathic and intranasal surgery was completed (Le Fort I, Sagittal ramus osteotomies of the mandible, genioplasty, septoplasty, reduction of inferior turbinates, removal of wisdom teeth). **a, b** Facial views at birth. **c, d** Facial views prior to and 1 year after jaw surgery. **e, f** Profile views before and 1 year after jaw surgery. **g–j** Occlusal views before and after reconstruction. **k, l** Lateral cephalometric radiographs before and 1 year after reconstruction

26.4 Residual Deformities

Ideally, the cleft patient will grow into adolescence without requiring additional procedures. The prevalence of residual deformities that require further surgical intervention vary considerably, depending on the cleft team's philosophy with regard to the staging of reconstruction and available surgical expertise. In addition, despite a team's preferred method of management in infancy, childhood, and early adolescence, a subgroup of cleft patients will present in adolescence with residual clefting problems including: maxillary hypoplasia, residual oro-nasal fistula, residual bony defects, cleft-dental gap(s), chin dysplasia/asymmetry, mandibular dysplasia/asymmetry, obstructive nasal breathing (septal deformity/enlarged inferior turbinates), and/or dental anomalies/partial anodontia [35–37].

26.5 Historical Perspective and Results

Over the decades, the literature has warned of the possible complications of maxillary osteotomy in the cleft patient but provided only limited descriptions of technique to guide the orthognathic surgeon in the performance of safe, reliable osteotomies to solve these problems [38–47]. In 1974, Willmar [48] reported on complications of 17 UCLP patients undergoing a LeFort I osteotomy. One patient had aseptic necrosis and partial loss of the lesser segment of the maxilla. In 1974, Georgiade [49] suggested that a camouflage approach with mandibular osteotomy and setback was often preferred to direct maxillary surgery. Kiehn and others [50], and Des Prez and Kiehn [51] warned of blood supply problems that might occur with maxillary surgery in cleft lip and palate patients. In 1975, Henderson and Jackson [52] reported combining lip-scar revision, anterior fistula closure, and maxillary osteotomy in a one-stage procedure. Their concept was innovative, but they did not specify the details of

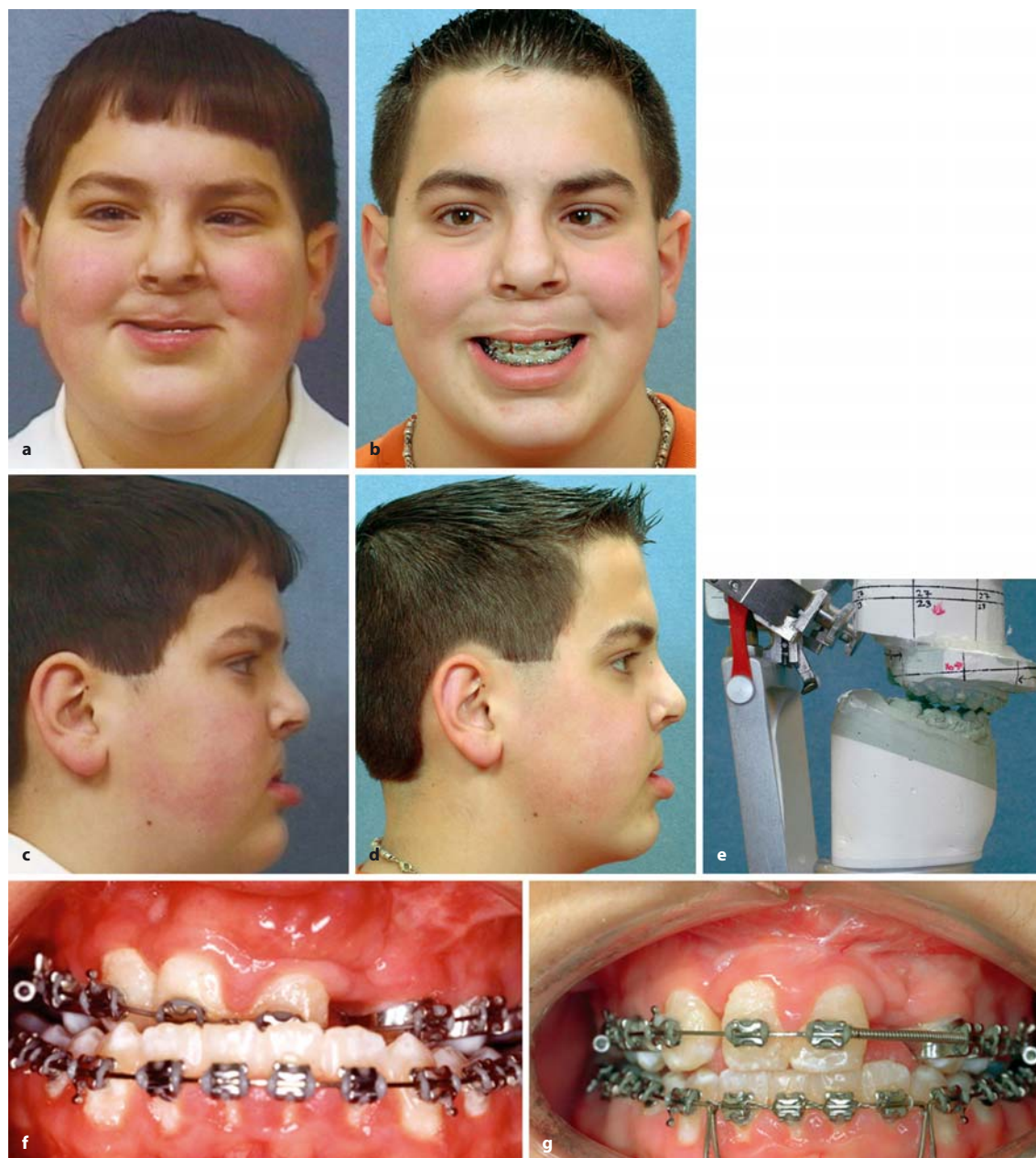


Fig. 26.2 a–i. 13-year-old boy born with BCLP. He was referred to the senior author after successful bone grafting but with maxillary hypoplasia. For psycho-social reasons “early” jaw surgery including a classic Le Fort I osteotomy with iliac crest bone

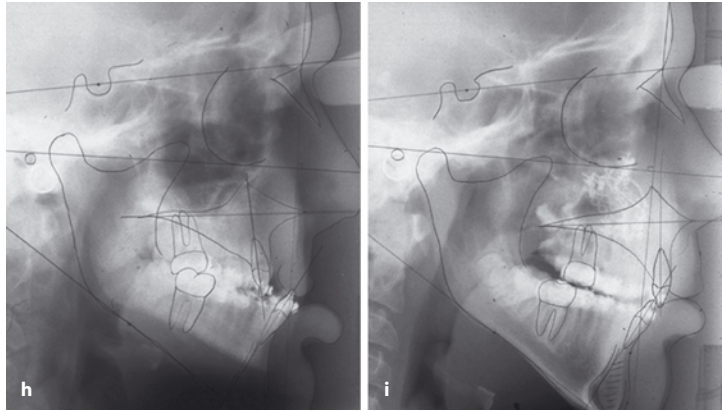
grafting was completed. **a, b** Facial views before and after LeFort I advancement. **c, d** Profile views before and after LeFort I advancement. **e** Articulated dental casts after model surgery. **f, g** Occlusal views before and after reconstruction

the technique. In 1978, Jackson [53] further described the LeFort I procedure as it applied to the cleft lip and palate patient, stating that if a large fistula was present that required extensive flap mobilization for closure, the maxillary blood supply might be in danger. In

such cases, he preferred a staged reconstruction with fistula closure and bone grafting followed later by osteotomy.

In general, surgeons have been leery of flap necrosis with loss of maxillary bone and teeth.

Fig. 26.2 a–i. (continued) 13-year-old boy born with BCLP. He was referred to the senior author after successful bone grafting but with maxillary hypoplasia. For psychosocial reasons “early” jaw surgery including a classic Le Fort I osteotomy with iliac crest bone grafting was completed. **a, b** Facial views before and after LeFort I advancement. **c, d** Profile views before and after LeFort I advancement. **e** Articulated dental casts after model surgery. **f, g** Occlusal views before and after reconstruction. **h, i** Cephalometric views before and after LeFort I advancement



In 1980, Tideman and coworkers [54] further refined the maxillary LeFort I osteotomy in cleft lip and palate patients with a segmental palatal osteotomy. Their incision placement requires significant subperiosteal degloving to avoid flap circulation risk but does not allow direct exposure. In 1980, Sinn [55] also reported on the simultaneous maxillary advancement, oronasal fistula repair, and bone grafting of the alveolar cleft. He stressed the importance of preserving a vertical soft tissue pedicle similar to that described by Tideman and associates. He then completed bony osteotomies through tunnels rather than under direct vision and used a cheek rotation flap for oral-side closure of the labial and palatal oronasal fistulas. With his technique, nonkeratinized buccal mucosa, rather than attached gingiva, is brought into the cleft tooth-bearing region. In 1984, Ward-Booth and colleagues [56] analyzed the LeFort II osteotomy used in their institution in 13 cleft patients for managing midface hypoplasia. They believed that the LeFort II osteotomy was preferable to the LeFort I osteotomy in providing an improved blood supply to the alveolar segments. The residual fistulas were not simultaneously closed and fixation was with direct wires, intermaxillary fixation (IMF), and craniomaxillary external appliances. In 1985, James and Brook [57] described a variation for the correction of maxillary hypoplasia in patients with cleft lip and palate by transection of the hard palate. They raised an extensive palatal flap and used three vertical stab incisions for exposure on the labial aspects of the maxilla. Access was then achieved through subperiosteal tunneling without direct exposure. Fixation was with IMF and a halo head frame, which was maintained for 10 weeks. The second procedure was required for further bone grafting and palatal fistula closure. The authors expressed concern that a more direct downfracture of the maxilla would risk vascular compromise to the segments. In 1985, Poole and others [58] proposed an additional modification of the Le Fort I

osteotomy to be used in patients with cleft palate. With their technique, a partial-thickness palatal flap was elevated, leaving the greater palatine vessels in situ and allowing the maxilla to be repositioned anteriorly without displacement of the soft palate. Poole and coworkers believed this approach would limit interference with velopharyngeal function. They again used small vertical incisions on the labial aspect, requiring tunneling and lacking direct exposure for osteotomies, disimpaction, fistula closure, bone graft placement, or plate-and-screw fixation. Fixation was with IMF, direct wires, and halo craniomaxillofacial fixation in this series.

For those patients who were either not bone grafted at all or in whom the graft did not take sufficiently to accomplish all objectives (adequate bone volume, fistula closure and/or eruption of the canine tooth) Posnick described modifications of the Le Fort I osteotomy to resolve these problems. Using this technique a successful outcome can be achieved by placing the soft tissue incisions to permit direct exposure for dissection, LeFort I segmental osteotomies, disimpaction, fistula closure, bone grafting, and application of plate-and-screw fixation but without risk of circulation injury to the dento-osseous-musculo-mucosal flaps [35, 59–63]. The increased visibility provided by these incisions makes possible the incorporation of the routine surgical closure of the cleft-dental gap through differential maxillary segmental repositioning. This method of approximating the cleft gap in the maxillary segments also closes the cleft dead space and brings together the labial and palatal mucosal flaps to permit closure of recalcitrant oronasal fistulas without tension (Fig. 26.3).

For the unsuccessfully bone-grafted BCLP patient who presents in adolescence with a cleft-jaw deformity, Posnick et al. also described methods for management [35, 59–65]. The most important variation from the UCLP technique (described above) is the placement of the incisions. The buccal incisions are made



Fig. 26.3 a–f. A 21-year-old born with UCLP was referred to the senior author to address his residual deformities: maxillary/mandibular/chin dysplasia, skeletal defects, oro-nasal fistula, and a cleft dental gap. He underwent a combined orthodontic and orthognathic surgical approach including: modified Le Fort I osteotomy in 2 segments; sagittal ramus osteotomies of

mandible; and genioplasty. A septoplasty and reduction of inferior turbinates was simultaneously carried out to improve nasal airflow. **a, b** Facial views before and 2 years after surgery. **c, d** Profile views before and 2 years after surgery. **e, f** Articulated dental casts after model surgery



Fig. 26.3 a-l. (continued) **g-l** Occlusal views before and after reconstruction

in the depth of the vestibule and extending from the zygomatic buttress and then forward to the location of the residual labial oronasal fistula on each side; the incisions are then taken down the mesial line angle of the canine teeth, (or if the canine is missing, the most mesial tooth in each lateral segment). These incisions separate the oral and nasal mucosa along each lateral segment. The premaxillary segment labial incisions are placed at the distal line angle of the central incisor on each side to separate the oral and nasal mucosa. Care is required to prevent any disruption of or incision into the mucosa on the labial vestibule

of the premaxilla. The nasal and oral mucosa are also sharply incised and separated on the palatal aspect of the premaxilla and on each lateral segment [35, 59–65].

Posnick and Thompson [62] prospectively assessed the presenting deformity and clinical results in 126 consecutive adolescents born with: UCLP ($n=66$); BCLP ($n=33$); and ICP ($n=17$) who underwent orthognathic surgery (by the same surgeon) requiring Le Fort I osteotomy. Sixty-one (92%) of the 66 UCLP patients underwent a modified LeFort I osteotomy and successful simultaneous oro-nasal fistula closure.

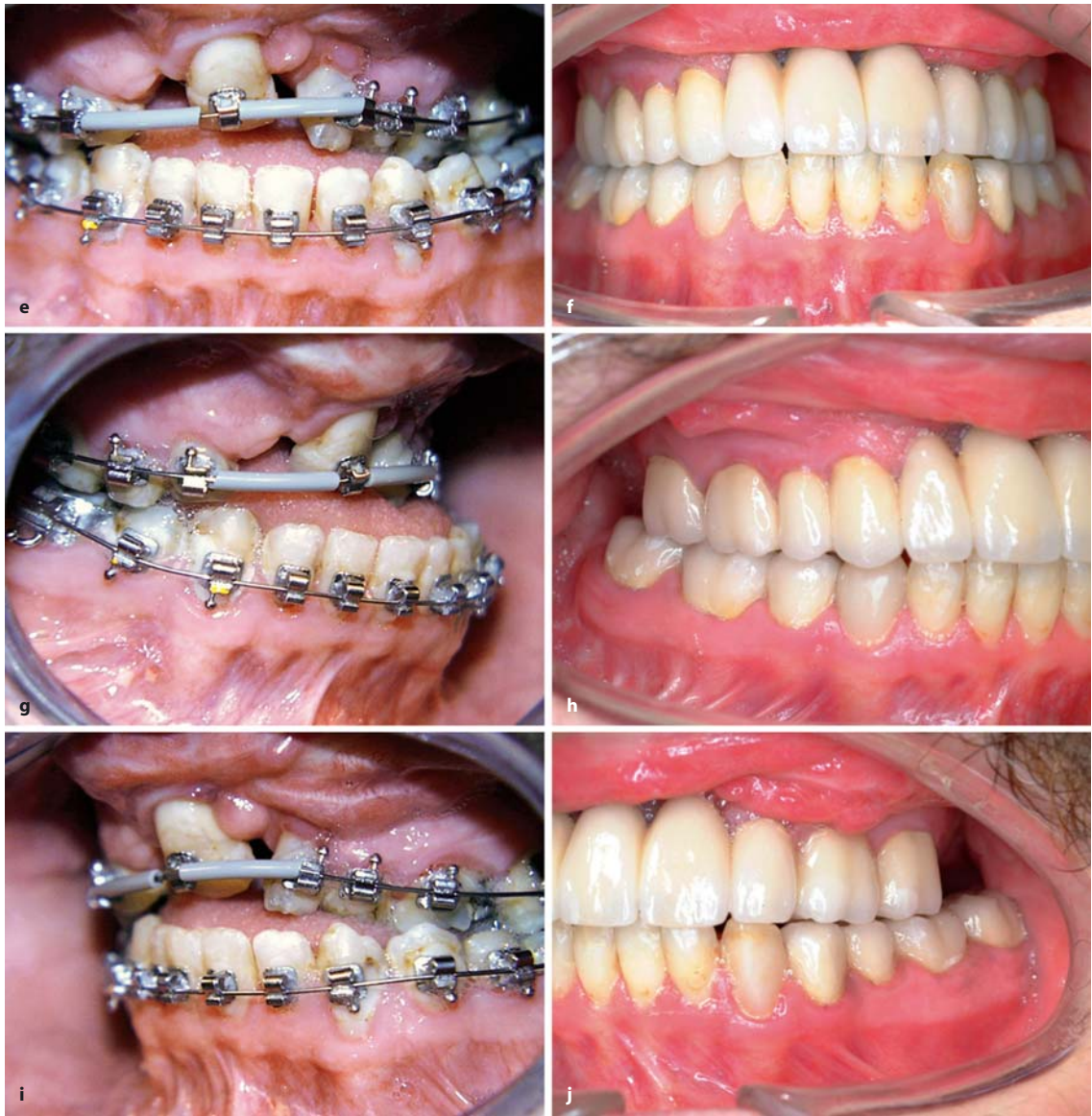


Fig. 26.4a-j. (continued) **e-j** Occlusal views before and 1 year after surgery/dental rehabilitation

able opportunity for the resolution of residual problems (i.e., maxillary hypoplasia, alveolar defects, residual fistulas, and cleft-dental gaps) [59].

Prosthetic Options: In the cleft patient, prosthetic options for the management of cleft-dental gaps exist. From a facial aesthetic, dental health, and speech perspective, the long-term use of a removable partial denture is always a second choice. Fixed bridgework is a viable alternative but requires the partial destruction of adjacent normal teeth, may look artificial,

requires replacement at intervals throughout the patient's life, and demands ongoing meticulous oral hygiene. In our opinion, these options should be reserved for the patient with a mutilated dentition (i.e., multiple missing and deformed teeth) (Fig. 26.4). Placement of a single-tooth osseous integrated implant is an attractive alternative for the missing lateral incisor at the cleft site, but the implant requires adequate bone height and volume, and quality keratinized soft tissue that is not routinely present [22, 26, 74–83]. In addition, it requires staged surgical proce-

dures, skilled prosthetic rehabilitation, is expensive, and may fail or look unnatural in the long run. Other dental refinements for a cleft patient's dysmorphic/hypoplastic/decayed anterior maxillary teeth include porcelain-veneer buildups, composite bonding, sculpting teeth, and bleaching techniques. Each of these refinements should be individualized and may improve the patient's dental aesthetics, smile, and self-esteem.

Velopharyngeal Dysfunction: Uncertainties about velopharyngeal function and management of an in-place pharyngeal flap should no longer be limiting factors when orthognathic surgery is necessary in a patient with a cleft. A nasoendoscopic guided clinical examination by a speech pathologist familiar with cleft palate and jaw deformities can reliably predict current and expected velopharyngeal function [26]. When significant postoperative velopharyngeal deterioration is anticipated, the patient and family are educated about the sequencing of treatment, and alternatives are discussed. Despite the frequent need for maxillary advancement to normalize the cleft patient's skeleton and facial aesthetics, the authors have not had to transect an in-placed pharyngeal flap to achieve the desired advancement. The hope that VPI could be prevented by using the "DO Technique" after Le Fort I osteotomy has not been realized.

Orthodontics: Due to the clefts effect on the developing dentition, virtually all individuals born with clefts through the alveolar ridge will require orthodontic intervention. If the alveolar process was adequately bone grafted in the mixed dentition, canine tooth eruption through the cleft site will take place. The lateral incisor is frequently anomalous or missing, and the adjacent teeth are typically rotated and tipped. Supernumerary teeth are common, but rarely is there room in the arch to accommodate them. If there is no orthognathic deformity then routine orthodontics is carried out. Camouflage orthodontics may be possible to "neutralize" the occlusion if the skeletal discrepancy is minimal, and if residual cleft defects (cleft dental gap, bone deficits, oronasal fistula) do not require further management. When a jaw deformity is present, orthodontics is carried out in conjunction with cleft-orthognathic surgery.

Distraction Osteogenesis: The option of distraction osteogenesis (DO) is advocated by some as the "technique of choice" for the cleft patient with maxillary hypoplasia [84]. Over the years, hoped-for advantages of the DO technique have included: "minimally invasive," "no pain," "minimal morbidity," "ability to treat earlier in life," "no VPI," "histogenesis of soft tissues," and "no relapse" [85]. Analysis of these hoped-for ad-

vantages to date has shown that the classic Le Fort I osteotomy approach with immediate repositioning remains the gold standard of treatment [86]. Since both techniques (classic and DO) are surgical approaches, they both carry the risks inherent with a general anesthetic and osteotomies: infection, airway difficulties, hemorrhage, nonunion, malunion, facial sensory loss, periodontal problems, devitalization of bone segments and teeth. Complications of cleft-orthognathic surgery using classic techniques are documented and in skilled hands occur infrequently [62]. The complications reported even by skilled surgeons using DO in conjunction with LeFort I osteotomy in cleft patients are of some concern [87]. The available DO devices continue to evolve, indicating unsatisfactory results with each previous generation of instrumentation used. In addition, many DO managed-cleft patients are treated for prolonged durations – with multiple "adjustments" required in the operating room and office – and with less-than-ideal results. With DO it is difficult to critically analyze specific "relapse" parameters because there is no set "finishing" point. Patients frequently continue treatment while the surgeon tries to "get it right" and to "allow consolidation" with the potential of dental compensation introduced due to excessive orthodontic traction. Questions have been raised as to the ethics and the scientific decision-making of clinicians when using these techniques [88, 89].

Using classic techniques in the cleft patient, careful preoperative planning and meticulous execution of the procedures is required and generally achieves the anticipated results immediately. The patient is more comfortable when no external appliances are required after surgery. No significant scalp or facial scars are produced. Postoperative participation by the patient and family is minimal because there are no office procedures or technical maneuvers to accomplish.

Other residual problems that the cleft patient may present with including: oronasal fistula; cleft dental gap(s); bone defects of the alveolar ridge/hard palate, floor of nose; transverse maxillary deficiency; mandibular disproportion and asymmetry; chin disproportion; dental anomalies; septal and turbinate abnormalities – can all be surgically managed simultaneously when using the classic orthognathic approach [86]. The patient can plan the surgery around their school, and/or work schedules as recovery time is predictable and relatively short. In general, these combined procedures are not possible when using the DO technique.

We agree with the opinion of Precious [90] when he states: "Conventional (classic) orthognathic surgery for the midface is safe, predictable, accurate and efficient. Treatment planning is reliable and duration of treatment is relatively short. Rigid internal

fixation is 'patient friendly' and a second surgical procedure to remove hardware is usually not necessary. Therefore at the present time traditional orthognathic surgical management remains the treatment of choice" [11].

Timing of Surgery: When asking the question – How early can you operate? – the technique used (DO vs. classic) does not make the difference. In fact, jaw surgery using either technique (DO vs. classic) can be done at virtually any age. The unknown variable of – *how much remaining mandibular growth is yet to occur?* – should be the determining factor when choosing the age of the patient at *definitive* jaw surgery. Neither technique (DO vs. classic) in conjunction with a Le Fort I osteotomy offers an advantage to: reducing maxillary scarring, stimulating/allowing further maxillary growth, or preventing further growth of the mandible.

26.7 Conclusions

For the cleft patient presenting in adolescence with a jaw discrepancy and malocclusion, misinformation and limited available surgical and dental expertise often prevents a favorable facial reconstruction and dental rehabilitation. A major advantage of the modified Le Fort I osteotomy is its ability to simultaneously: close cleft dental gap(s), resolve oro-nasal fistulas, manage skeletal defects, stabilize dento-alveolar segments, and correct jaw deformities. When a thoughtful staging of reconstruction is undertaken, individuals born with cleft lip and palate can reach adolescence after undergoing only a limited number of operations and interventions, without negative attention being drawn to their original malformation.

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27 Prevention of Relapse Following Cleftal Bone Grafting and the Future Use of BMP Cytokines to Regenerate Osseous Clefts Without Grafting

PHILIP J. BOYNE, ALAN S. HERFORD, DALE E. STRINGER

The problem of possible interference with the growth of the premaxilla in cleft patients and/or relapse of the maxilla following orthodontic therapy, and following surgical bone grafting, is a very serious one which has been the subject of much clinical research and debate during the past four decades. A discussion of the problem can be approached by dividing the areas of concern into: (a) proper sequencing of bone grafting and orthodontic treatment and (b) the prevention of relapse following secondary bone grafting and following orthognathic surgery in the older age groups.

Historically, the present popular use of bone grafting in the form of particulate cancellous bone from the iliac crest was developed by Boyne in 1970 [1–3]. The adoption of the use of particulate marrow and cancellous bone grafting system (PMCB) from the ilium followed a period of de-emphasis of bone grafting after the employment of rib grafts and other types of cortical and cancellous graft materials had resulted in many reported cases of delay in osseous growth of the premaxilla. The poor long-term results from use of rib grafts diminished the surgeons' interest in bone grafting for approximately a decade prior to graft resurgence in 1970 [4, 5]. It was found that PMCB grafting (Fig. 27.1) resulted in restoration of height and width of the defect. Furthermore, the cancellous bone in the bone grafted area could be stimulated by eruption of the canine tooth when the patient being grafted was around the age of 8 through 11. Thus the postgraft stimulation of the cleft and stimulation of osseous maturation through remodeling of the graft was provided primarily by natural or orthodontic tooth eruption into, adjacent to, the grafted area [6, 7].

Subsequently, it was found that bone grafting in earlier age groups could be successful, provided the graft could be appropriately stimulated by orthodontic therapy or orthopedic forces following the grafting procedure [8, 9].

This chapter reviews clinical results and problem areas involved with grafting and analyzes possible



Fig. 27.1. View of the type of autogenous cancellous bone and marrow (PMCB) utilized in cleft palate grafting. The graft has been taken from the anterior iliac crest

causes for relapse, malocclusion, and other clinical sequelae that may be seen in completing the treatment of these patients following osseous grafting procedures.

Additionally, the application of tissue engineering concepts have expanded the possibility of osseous regeneration of the bony cleftal defect without actually using bone grafts of any type.

The problem of prevention of long-term complications following bone grafting to maxillary anterior clefts is usually the result of the application of surgical and orthopedic techniques which have inherent design deficiencies which tend to lead to unfavorable results. Extended observations of the results of certain types of grafting and orthopedic procedures should lead the clinician to adapt a treatment concept which is optimal for the child patient. Unfortunately, in the past, grafting procedures and surgical para-grafting techniques have been undertaken and utilized for a long period of time without a careful review of the long-term results.

It is imperative, for example, that in osseous grafting of unilateral clefts, a procedure be undertaken that produces more than a simple tethering or a union of the lateral bony segmental margin of the cleft with the anterior premaxillary segment. Rather, the grafting surgery should produce an optimal thickness of new bone in the labial-palatal dimension of the cleftal area. This maximal amount of bone mass is necessary to produce a base for eruption of the canine tooth, or the maintenance of the lateral incisor, if present, and the proper positioning of the adjacent central incisor. Additionally, the graft should produce sufficient bone to aid in the maintenance of osseous support in the orthodontic eruption and movement of the posterior teeth.

Also the optimal amount of bone in the grafted reconstructed area should be of the type to respond well to maxillary growth, as well to orthopedic forces of orthodontic movement in expansion of the arch to maintain the patient in a *proper noncrossbite occlusion*.

The technique of using a periosteoplasty within the first year of the patient's life together with the insertion of a small cortical bone graft consisting of a thin bony graft matrix produces a union (in a unilateral cleft) of the short alveolar segment with the large premaxillary and contralateral segment. However, the union is capricious and insufficient to actually maintain the arches and to resist *crossbite development*. This unfavorable long-term result is related to the graft producing an insufficient bony matrix base referred to above. Thus, it is our view that the periosteoplasty procedure (Rosenstein and others) [10] has not

been appropriately evaluated over a long period of time by critical review of these important specific problem areas.

It is felt that to produce optimal long-term results, it is far better to use early secondary bone grafting at the ages of 4 or 5 to 6 as advocated by Precious and others [8, 9].

27.1 Iliac Crest Bone Grafting

We have been using iliac crest bone grafts for the past 25 years with excellent success provided the proper regimen and coordination of orthodontic treatment is followed and provided sequencing of surgery and orthodontic treatment is appropriate.

In cases treated with this protocol, patients do not tend to experience relapse, lack of growth, malocclusion, asymmetry, and other clinical problems. As can be seen in Table 27.1, we have found that grafting at earlier ages (5–7 years) gives excellent results when appropriate scheduling of therapy is followed [7, 8]. Table 1 shows that PMCB grafting in the 5- to 7-year-old age group followed by proper positioning of the central incisors, rotating the central incisor from the 90° torque position, placing the central incisor in proper anteroposterior relationship to the mandibular incisors, and correcting the maxillary midline appropriately stimulates the graft so that the proper critical mass of osseous material is available for the orthodontist to complete the treatment and move the teeth in completion of the dental arch.

Table 27.1. Sequencing of treatment in cleft palate patients

Age of Point	Orthodontic Rx	Surgical Rx
6–8 weeks		Cleft lip adhesion without passive molding plates
3–6 months		Cleft lip repair
2 1/2–3 years		Velopharyngeal surgery if needed
5–7 years	After bone grafting: maxillary alignment of incisors into grafted area Horizontal expansion A-P expansion	The most opportune time for: PMBC graft, and closure of oronasal fistula
7–8 years	Eruption of maxilla centrals into alignment Orthodontic tooth eruption Guidance and positioning Completion of transverse posterior expansion	
9–11 years	Arch alignment Tooth positioning Cuspid eruption into the grafted area	
12–14 years	Orthodontic retention	
14–15 years	Final arch alignment	
15–20 years		Orthognathic surgery if necessary Nose and lip revision

When there are lapses in the treatment due to non-compliance of the patient or conflicts in treatment plan, the graft material does not undergo appropriate proliferation, maturation, and remodeling. There is an incomplete bridging of the cleft, usually with only a labial osseous bridge with insufficient bulk of cancellous bone from the labial portion palatally, so that movement and positioning of the central incisors and canine teeth into the area at a later age result in the placement of the roots of these teeth into a void or space with insufficient bone around the roots which tends to lead to instability, relapse, loss of teeth, root resorption, and other problems.

27.2 Appropriate Sequencing of Orthodontic and Surgical Treatment

Both bilateral and unilateral clefts have the potential for developing relapse. However, the bilateral cleft has greater tendency in this regard; and the possibility of persistent malocclusion and untoward postoperative sequelae is greater in the bilateral clefts. It has been our experience since beginning iliac crest PMCB grafting with bilateral and unilateral clefts in 1969 that appropriate stimulation of the graft postsurgically is extremely important. The most effective stimulation of the graft causing proliferation of bone and appropriate remodeling is through tooth movement and arch expansion. One of the presenting problems which occurs early in both unilateral and bilateral clefts is the anteroposterior malpositioning of the incisor teeth. (Figs. 27.2–27.5) If the anteroposterior position of the incisor teeth is not corrected, the lingual “lock” of the incisor or incisors will further inhibit the development of the maxilla. If there is proper incisor relationship, the maxilla will grow appropriately.

Enlow [11] has shown that the premaxilla will grow downward with appropriate positioning by the cooperative interaction of bone resorptive areas and bone deposition on the periosteal surface of the anterior maxilla as well as by sutural growth. As the mixed dentition stage proceeds to the permanent dentition the alveolar process itself tends to bring the premaxilla into its correct relationship, provided that the permanent maxillary incisors have been positioned correctly and are supported by a sufficient osseous base produced by acceptable bone grafting (Figs. 27.6–27.8). Both of these growth actions are necessary for the proper relationship of the premaxilla. Once correct anatomic form has been established, the tendency for relapse and postoperative complication is greatly reduced.



Fig. 27.2. View of torsion of the maxillary central incisor on the left side with tendency toward incisor malpositioning and maxillary anteroposterior osseous deficiency

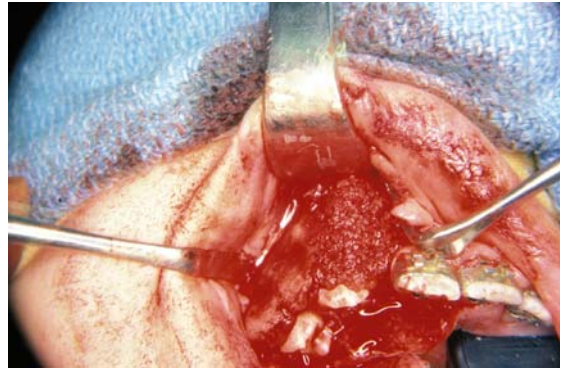


Fig. 27.3. Intraoperative view showing placement of the particulate cancellous bone and marrow in the cleft area after careful closure of the nasal soft tissue floor. This type of graft provides an osseous base for correction of the torsion of the central incisor and a base for movement of the central incisor labially out of AP crossbite position



Fig. 27.4. A view of an appliance used to stabilize the posterior occlusion and to maintain an anterior open bite while the central incisor is brought forward from its “blocked” lingual crossbite position to the proper incisal relationship with the lower incisor teeth after (PMCB) bone grafting



Fig. 27.5. View after the central incisor and lateral incisors have been brought out of torsion and placed in the proper AP and transverse relationship. The canine tooth has also been erupted into position. (The lateral incisor also may be salvaged in this manner in approximately 10% of cases.)



Fig. 27.7. A lateral view of the patient in Fig. 27.6 showing the extent of the large oronasal fistulas. After grafting, the posterior arch was expanded and the arch was brought into proper alignment and the canine tooth was erupted into position as shown in Fig. 27.8



Fig. 27.6. A view of a bilateral cleft in the mixed dentition stage prior to grafting showing large oronasal fistulas and "downward positioning" of the premaxilla



Fig. 27.8. Completed case shown in Figs. 27.6, 27.7 with excellent arch form and with the canines having been brought orthodontically to the central incisors

Delays in stimulation of the PMCB graft of the premaxilla by orthodontic treatment can lead to serious postoperative problems. As previously stated, the most favorable stimulation of the graft is by arch expansion or by movement of the teeth, particularly the canine tooth, if the age is appropriate, or by moving the central incisors out of anteroposterior crossbite in the younger age groups. Where there is no stimulation of the graft, there tends to be a slowing of the anteroposterior growth which tends to produce the sequelae of impacted canine teeth, "locking" or lingual collapse of the maxillary central incisors, and collapse of the premaxilla arch, which subsequently is resistant to expansion and usually requires surgical osteotomy for correction of the deficit. These sequelae have been observed when arch expansion has been delayed and instituted late, 1–2 years after grafting. We therefore recommend that orthodontic stimulation be institut-

ed within a period of 6 months after PMCB grafting in all cases in which a anteroposterior crossbite or a transverse posterior crossbite and malocclusion exist.

Over the past 30 years, we have found that by offering bone grafting to patients of 5–7 years of age when the central incisors are presenting in torsion and in crossbite, orthodontic treatment may be instituted immediately after grafting (i.e., 12 weeks) to bring the anterior teeth out of crossbite position (Figs. 27.9, 27.10). This system of treatment tends to avoid horizontal hypoplasia of the maxilla, thus the patients have a shorter period of orthodontic treatment [6–8] and do not require orthognathic surgery at a later date (Fig. 27.11). This combined interceptive bone grafting and orthodontic treatment at an early age avoids more extensive prolonged treatment later in the patient's life.

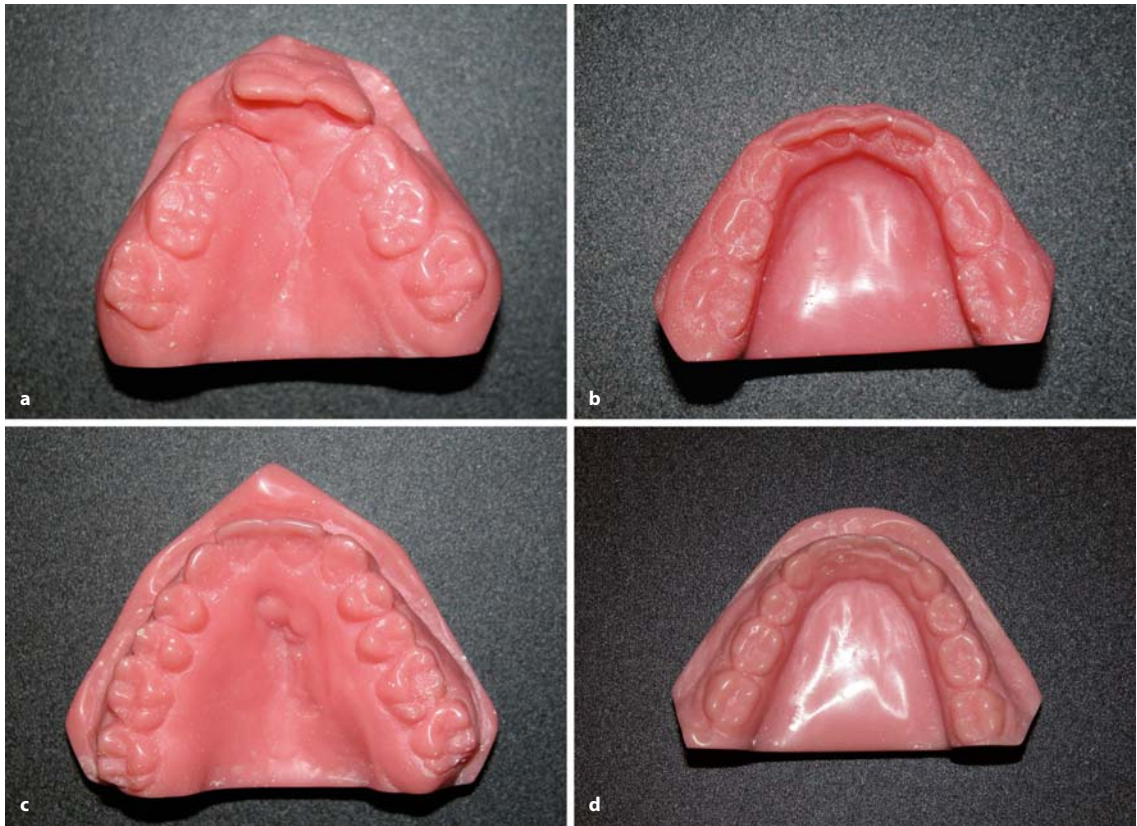


Fig. 27.9. **a** Occlusal maxillary views of models of a bilateral cleft case in the mixed dentition stage with posterior transverse collapse leading to crossbite. The patient was grafted early at the age of 7, the arch expanded, and the canine teeth moved into

appropriate positions. **b** Mandibular arch preoperatively. **c** Occlusal views of maxillary arch after PCMB grafting and orthodontic alignment. **d** Occlusal view of mandibular arch after treatment

There has been reluctance by some orthodontists at times to institute treatment early in cleft patients. This has been based on the view that (a) there tends to be poor compliance by patients of this age, (b) the maxilla when grafted at an early age may exhibit arresting of vertical and anterior maxillary growth and development, and (c) the overall orthodontic treatment time may be prolonged leading to future patient noncompliance. However, it has been our experience in grafting children between the ages of 5 to 7 years old that the majority are cooperative and compliant and that the necessary orthodontic treatment can be undertaken at that time. This experience has been shared by others [9].

Semb [12], in reporting on over 562 cases, noted that 25% were treated by orthodontic appliances to correct the developing or existing anteroposterior

crossbite at ages 6–7. In cases treated early, the pre-maxilla was stabilized in proper occlusion and the patient monitored until the age of 9–11 when the canine tooth was erupted. Semb did not report any problem with patient compliance [8].

In our series of cases, we found that orthodontic treatment time using early (5–7 years of age) bone grafting protocols was decreased rather than increased. Therefore, the opportunity for patient “burnout” is less.

There have been no reported detrimental effects on osseous growth, arch development, or arch alignment in large groups of patients treated by cleft palate bone grafting in Europe using our technique. A delay in establishing proper positioning of the maxillary incisor teeth tends to produce problems because of the lack of orthodontic stimulation of the graft site.

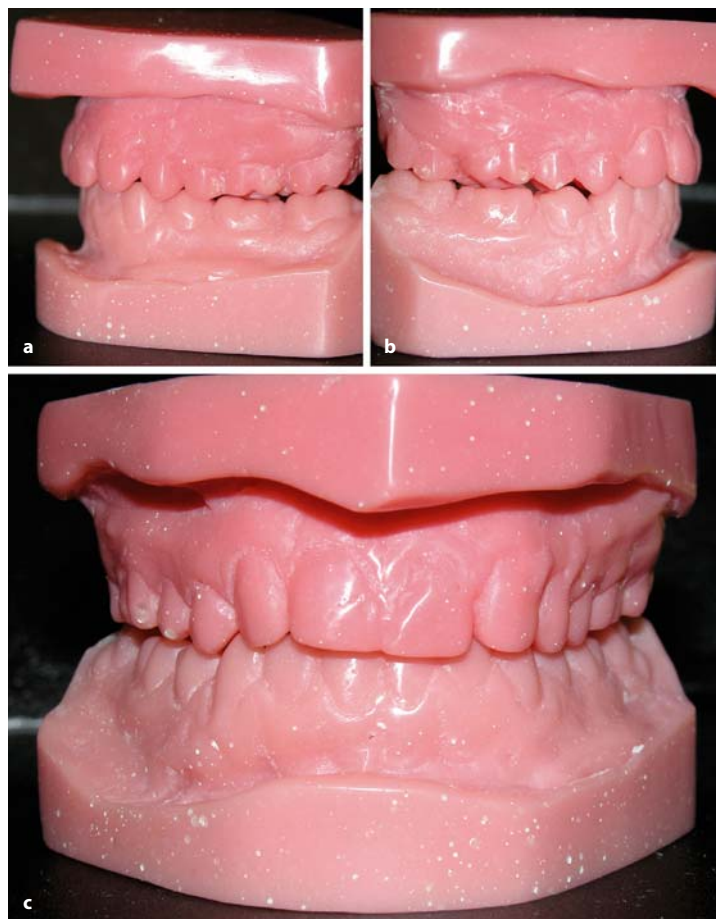


Fig. 27.10 a–c. Views of the completed case shown in Fig. 27.9 with the patient completely out of retainers at age 14. **a** Left lateral view. **b** Right lateral view. **c** Frontal view



Fig. 27.11. At times, the canine tooth may be moved laterally producing an edentulous space in which there is ample bone formation due to the proper placement of the graft. Excellent bone is present for placement of a rootform implant or for the placement of a fixed prosthesis

27.3 Description of the Surgical Procedure for Bone Grafting

This surgical procedure produces a necessary bulk of bone graft material in the area of the cleft extending palatally from the labial plate at least a centimeter along the horizontal palate. A sufficient bulk of bone must be maintained and established in this area to stabilize the alveolar process (Fig. 27.11). A bone graft placed only on the labial portion tends to remodel to produce a cortical plate, but very little cancellous bone in the alveolar ridge area itself. This makes it very difficult for the orthodontist to move teeth into the area. In addition, periodontal deficiencies occur as the canine tooth is moved into alignment and as the central incisors are brought out of the torsion position. In order to produce an osseous base in the area through the use of graft material adequately placed on the palatal side of the alveolar ridge area, it is necessary that palatal flaps be raised (Figs. 27.12–27.15). If this is not done, the graft material is restricted to the area along

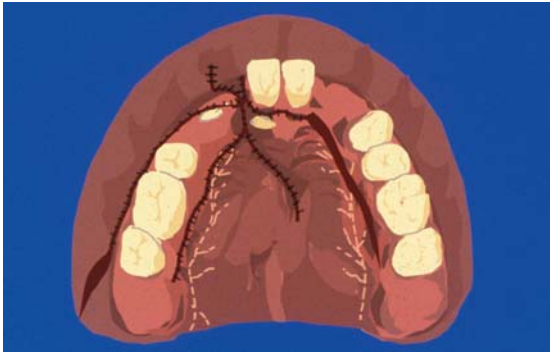


Fig. 27.12. a drawing showing the outline of the flaps with the blood supply from the greater palatine artery to the soft tissue which will be moved to close the palatal portion of the fistula. Labially, the posteriorly based buccal flap, which has ample blood supply, is moved anteriorly and extended over the crest of the ridge to cover the bulk of the graft (as in Figs. 27.14, 27.16). In this manner, sufficient graft material is placed for an adequate alveolar osseous base



Fig. 27.13. Intraoperative view showing migration of the palatal flap to close the palatal fistula over the bone graft. The bone graft has been extended from the labial crest to the palatal aspect of the alveolar ridge

the labial portion of the cleft defect. Further, without a palatal flap, the surgeon cannot adequately close the nasal mucosa on palatal aspect of the alveolus; particles of the graft will therefore be extended into the floor of the nose and a postoperative oral nasal fistulae will remain. The stage is set for a palatal periodontal deficiency of alveolar bone involving the central incisor and the canine tooth.

Thus, if the graft material does not extend sufficiently toward the palate in the area of the alveolar ridge, there will be lack of cancellous bone and osseous instability will occur in the area which will militate against good maintenance of the expanded arches and of good functional occlusion (Fig. 27.16).

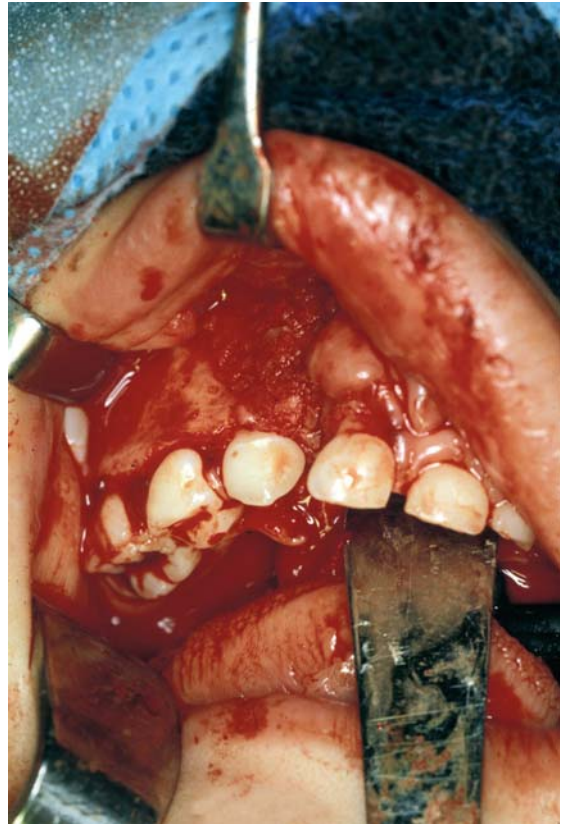


Fig. 27.14. The labial portion of the graft intraoperatively shows graft material being placed. The graft extends from labial to palatal aspects of the ridge, supplying an appropriate osseous base

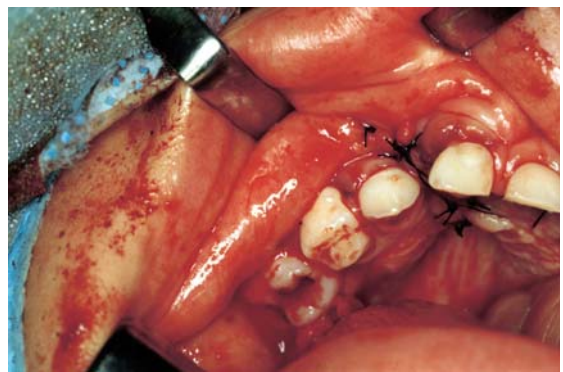


Fig. 27.15. An intraoperative view showing the closure of the palatal and labial flaps



Fig. 27.16. Another intraoperative view showing the posteriorly based labial flap which has been migrated over the occlusal portion of the ridge for complete soft tissue closure over the graft material

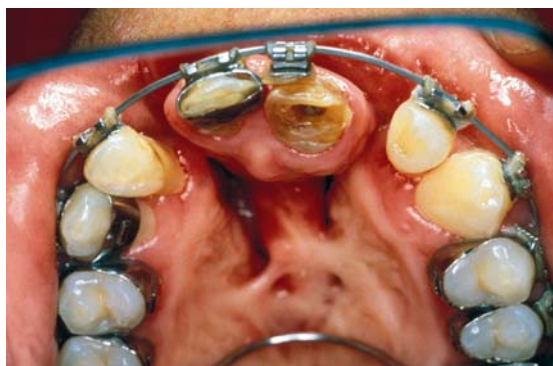


Fig. 27.17. A clinical case showing the result of prolonged manipulation of the premaxilla without prior grafting. The very narrow vomer stalk does not permit adequate stabilization of the premaxilla prior to grafting. Orthodontic movement prior to grafting leads to a widening of the defect, loss of bone of the premaxilla, and difficulty in establishing a bone graft to unite the premaxilla with the transversely expanded posteriorly maxillary base. Such prolonged treatment is another major factor in causing long-term maxillary relapse



Fig. 27.18 a–g.

27.4 Relapse in Bilateral Cases Following Bone Grafting and Possible Causes of Relapse

Relapse of the premaxilla, loss of orthodontic control of the premaxilla, and difficulty in maintaining midline tend to follow late bone grafting or prolonged orthodontic treatment prior to bone grafting. The unstable premaxilla with the small amount of maxillary bone attached to the vomer is usually incapable of being maintained with good stability without grafting. Therefore, long-term orthodontic manipulation of the maxilla as the patient continues to increase in age prior to grafting leads to marked instability of the premaxilla and difficulty in maintaining anteroposterior growth (Fig. 27.17). This situation results in the patient coming to orthognathic surgery needing a multiple-piece LeFort I osteotomy, which carries a high risk of postorthognathic surgical relapse because it is difficult to move the maxilla forward more than

5–7 mm even in conjunction “two-jaw” or bimaxillary surgery. This difficulty in moving the segmented maxilla and difficulty in maintenance of forward placement of the maxilla has been a major factor in relapse after orthognathic surgical treatment of cleft cases (Figs. 27.18, 27.19). When the premaxilla has been effectively grafted and the anteroposterior incisor relationship corrected, any need to move the maxilla forward at a later date can be accomplished by a single one-piece LeFort osteotomy with a much-diminished possibility of relapse.

Orthognathic surgical procedures, when necessary in bilateral cleft cases, can be successful with the use of rigid or semi-rigid fixation, with movement of a one-piece maxilla which has been previously grafted.

27.5 The “Displaced” Premaxilla

In certain bilateral clefts, there is a marked apparent “displacement” of the premaxilla anteriorly. Dealing with this defect from an osseous standpoint has been the subject of a great deal of conjecture.

Although some surgeons advocate a surgical fracturing of the premaxilla from the vomer “stalk,” with retrusion of the attached premaxilla, we feel that this procedure is not appropriate. A tremendous decrease in the growth potential of the premaxilla occurs when the septum is surgically fractured and repositioned.

The reason for the originally unfavorable premaxillary position is usually an early transverse collapse of the posterior arch. Appropriate treatment, therefore, involves expansion of the posterior arch, allowing the prepalate to assume its normal place in maxillary growth. Further, the premaxilla should be joined by bone grafting to the remaining posterior arch, enabling the entire maxilla to assume its normal growth pattern as a unit rather than as a multipiece structure (Figs. 27.20, 27.21).

The loss of the potential for downward and forward growth from the vomer stalk should be avoided [11]. The method of treatment for these cases therefore should be early grafting with PMCB taken from the

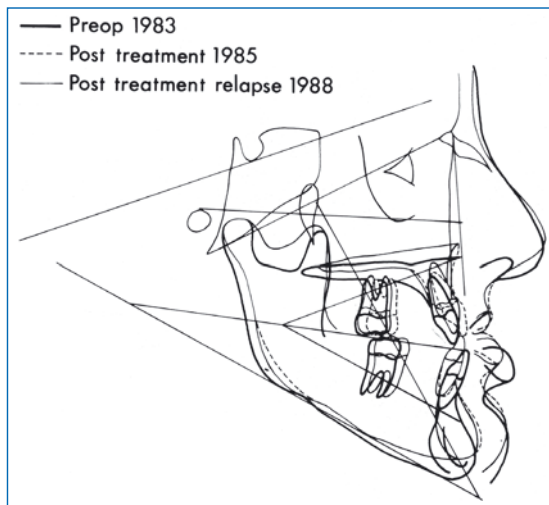


Fig. 27.19. A cephalometric tracing of the original postoperative position (solid line), a tracing 2 years after the orthognathic surgery (dashed line), and a third tracing 5 years (grey line) later showing the relapse which required reoperation

Fig. 27.18. **a** A profile view of a patient with cleft prepalate and posterior palate, a recessive maxilla, and a prognathic mandible. This patient required LeFort I osteotomy in conjunction with mandibular setback by bilateral sagittal split osteotomy. **b** Full-face view of the patient preoperatively. **c** An intraoperative view of the LeFort I procedure showing the cleft from the down-fracture position, demonstrating the width of the cleft which required bone grafting at the time of the movement of the maxilla. In a multisegmented LeFort I cleft maxilla of this

nature, the anterior movement is restricted to about 5–8 mm when made in conjunction with the posterior setback of the mandible. **d** A lateral view after surgery showing the establishment of a normal aesthetic profile to the face. **e** A lateral view 3 years postoperatively showing relapse to previously existed condition. **f** Final view of full face 3 years after reoperation. **g** Final lateral view after reoperation 5 years after the initial orthognathic surgical procedure. A LeFort I osteotomy was performed to establish normal Class I occlusion as seen here



Fig. 27.20. A bilateral cleft with large oronasal fistulas bilaterally and palatally with a “displaced premaxilla.” The premaxilla is not really “displaced,” but is in a normal vomer ethmoid relationship. The posterior maxillary arch has collapsed, requiring expansion with appropriate bone grafting to unite the primary palate with the secondary palate. The basis for a normal relationship can be established by this method of treatment



Fig. 27.21. A view of the excellent alignment of the teeth that can be established by the system described for bilateral cases presenting initially with a marked unfavorable anterior relationship of the premaxilla

iliac crest and rapid posterior arch expansion. The graft will “expand” by remodeling as the arch is enlarged. On the other hand, the ungrafted cleft defect will become even larger with the expansion of the posterior arch, making later bone grafting difficult. For this reason, we advocate grafting early before expansion. By approaching the bony clefts through a posteriorly based buccal flap bilaterally along with appropriate palatal flaps, the premaxilla can be grafted to both posterior maxillary segments.

Postoperatively, an appliance is inserted to expand the collapsed posterior arch transversely to its normal position. The prepalate then can be aligned with the posterior maxilla.

27.6 The Use of Different Types of Bone Graft Materials

Although different types of bone graft substitutes – xenogeneic, alloplastic and allogeneic – have been used at times in cleftal areas of growing children, we do not advocate such procedures, except in certain adults for alveolar augmentation of the cleftal area. In the adult patient, once an appropriate bridge of viable bone has been secured by an autogenous graft, a bone graft substitute (e.g., xenograft material such as Bio-oss) can be used to augment the existing osseous bridge. However, this type of a graft substitute is not recommended as a primary graft to obtain the initial osseous reconstruction of the cleft defect in growing children because such an osseous substitute material does not readily remodel to form viable bone to receive tooth movement and eruption. There is a tendency for normal growth to be restricted in the grafted area. Additionally, the grafted area will not respond with normal osseous repair phenomena when subjected to orthodontic treatment. Because large pieces of unremodeled residual bone substitute material (alloplastic particles and/or allogeneic particles) remain in the area, the normal osteoclastic-osteoblastic remodeling process is severely impaired.

The use of such materials in adult clefts, however, has a place to augment both the labial thickness and alveolar height to create a proper recipient site for the rootform implant. Allografts, xenografts and alloplastic materials may be used in adults having rootform implants placed in the cleftal areas.

27.7 Surgical Use of BMP Cytokines in Producing Appropriate Reconstruction of Cleftal Bone Defects

It is believed that the restoration of the osseous cleft to a full natural state without residual bony defect, with the former cleftal area having a normal thickness of the bone cortices and an appropriate thickness and distribution of an appropriate trabecular bone pattern should be the goal of all grafting procedures.

As mentioned in the introduction, unfortunately in the past osseous restoration procedures were undertaken using large segments of solid graft material that resulted over the long-term in residual areas of unremodeled cortical graft matrix in the surgical site. It is believed that this type of residual graft matrix does not contribute to the maxillary growth of the cleft palate patient and further may impede the orthodontic movement of teeth into the cleftal area. Therefore, graft techniques that restore the natural state of the bone as quickly as possible are considered to be pro-

CYTOKINE NOTES

SELECTED CYTOKINES AND CHEMOKINES

XVI

XI XII XII/XIII XIII XIV/XV

VEGF-A EGF HB-EGF IL-6 SCF

VEGF-B TGF-α SMDf LIF FIt-3L

AR IL-11 CT-1 M-CSF

BTC G-CSF CNTF MK

OB OSM PTN MIS

CyTocine Family Key

Chemokine Family Key

Key

TNF-α

Tumor Necrosis Factor Alpha

Receptors: α₁, α₂, α₃, α₄, α₅, α₆, α₇, α₈, α₉, α₁₀, α₁₁, α₁₂, α₁₃, α₁₄, α₁₅, α₁₆, α₁₇, α₁₈, α₁₉, α₂₀, α₂₁, α₂₂, α₂₃, α₂₄, α₂₅, α₂₆, α₂₇, α₂₈, α₂₉, α₃₀, α₃₁, α₃₂, α₃₃, α₃₄, α₃₅, α₃₆, α₃₇, α₃₈, α₃₉, α₄₀, α₄₁, α₄₂, α₄₃, α₄₄, α₄₅, α₄₆, α₄₇, α₄₈, α₄₉, α₅₀, α₅₁, α₅₂, α₅₃, α₅₄, α₅₅, α₅₆, α₅₇, α₅₈, α₅₉, α₆₀, α₆₁, α₆₂, α₆₃, α₆₄, α₆₅, α₆₆, α₆₇, α₆₈, α₆₉, α₇₀, α₇₁, α₇₂, α₇₃, α₇₄, α₇₅, α₇₆, α₇₇, α₇₈, α₇₉, α₈₀, α₈₁, α₈₂, α₈₃, α₈₄, α₈₅, α₈₆, α₈₇, α₈₈, α₈₉, α₉₀, α₉₁, α₉₂, α₉₃, α₉₄, α₉₅, α₉₆, α₉₇, α₉₈, α₉₉, α₁₀₀, α₁₀₁, α₁₀₂, α₁₀₃, α₁₀₄, α₁₀₅, α₁₀₆, α₁₀₇, α₁₀₈, α₁₀₉, α₁₁₀, α₁₁₁, α₁₁₂, α₁₁₃, α₁₁₄, α₁₁₅, α₁₁₆, α₁₁₇, α₁₁₈, α₁₁₉, α₁₂₀, α₁₂₁, α₁₂₂, α₁₂₃, α₁₂₄, α₁₂₅, α₁₂₆, α₁₂₇, α₁₂₈, α₁₂₉, α₁₃₀, α₁₃₁, α₁₃₂, α₁₃₃, α₁₃₄, α₁₃₅, α₁₃₆, α₁₃₇, α₁₃₈, α₁₃₉, α₁₄₀, α₁₄₁, α₁₄₂, α₁₄₃, α₁₄₄, α₁₄₅, α₁₄₆, α₁₄₇, α₁₄₈, α₁₄₉, α₁₅₀, α₁₅₁, α₁₅₂, α₁₅₃, α₁₅₄, α₁₅₅, α₁₅₆, α₁₅₇, α₁₅₈, α₁₅₉, α₁₆₀, α₁₆₁, α₁₆₂, α₁₆₃, α₁₆₄, α₁₆₅, α₁₆₆, α₁₆₇, α₁₆₈, α₁₆₉, α₁₇₀, α₁₇₁, α₁₇₂, α₁₇₃, α₁₇₄, α₁₇₅, α₁₇₆, α₁₇₇, α₁₇₈, α₁₇₉, α₁₈₀, α₁₈₁, α₁₈₂, α₁₈₃, α₁₈₄, α₁₈₅, α₁₈₆, α₁₈₇, α₁₈₈, α₁₈₉, α₁₉₀, α₁₉₁, α₁₉₂, α₁₉₃, α₁₉₄, α₁₉₅, α₁₉₆, α₁₉₇, α₁₉₈, α₁₉₉, α₂₀₀, α₂₀₁, α₂₀₂, α₂₀₃, α₂₀₄, α₂₀₅, α₂₀₆, α₂₀₇, α₂₀₈, α₂₀₉, α₂₁₀, α₂₁₁, α₂₁₂, α₂₁₃, α₂₁₄, α₂₁₅, α₂₁₆, α₂₁₇, α₂₁₈, α₂₁₉, α₂₂₀, α₂₂₁, α₂₂₂, α₂₂₃, α₂₂₄, α₂₂₅, α₂₂₆, α₂₂₇, α₂₂₈, α₂₂₉, α₂₃₀, α₂₃₁, α₂₃₂, α₂₃₃, α₂₃₄, α₂₃₅, α₂₃₆, α₂₃₇, α₂₃₈, α₂₃₉, α₂₄₀, α₂₄₁, α₂₄₂, α₂₄₃, α₂₄₄, α₂₄₅, α₂₄₆, α₂₄₇, α₂₄₈, α₂₄₉, α₂₅₀, α₂₅₁, α₂₅₂, α₂₅₃, α₂₅₄, α₂₅₅, α₂₅₆, α₂₅₇, α₂₅₈, α₂₅₉, α₂₆₀, α₂₆₁, α₂₆₂, α₂₆₃, α₂₆₄, α₂₆₅, α₂₆₆, α₂₆₇, α₂₆₈, α₂₆₉, α₂₇₀, α₂₇₁, α₂₇₂, α₂₇₃, α₂₇₄, α₂₇₅, α₂₇₆, α₂₇₇, α₂₇₈, α₂₇₉, α₂₈₀, α₂₈₁, α₂₈₂, α₂₈₃, α₂₈₄, α₂₈₅, α₂₈₆, α₂₈₇, α₂₈₈, α₂₈₉, α₂₉₀, α₂₉₁, α₂₉₂, α₂₉₃, α₂₉₄, α₂₉₅, α₂₉₆, α₂₉₇, α₂₉₈, α₂₉₉, α₃₀₀, α₃₀₁, α₃₀₂, α₃₀₃, α₃₀₄, α₃₀₅, α₃₀₆, α₃₀₇, α₃₀₈, α₃₀₉, α₃₁₀, α₃₁₁, α₃₁₂, α₃₁₃, α₃₁₄, α₃₁₅, α₃₁₆, α₃₁₇, α₃₁₈, α₃₁₉, α₃₂₀, α₃₂₁, α₃₂₂, α₃₂₃, α₃₂₄, α₃₂₅, α₃₂₆, α₃₂₇, α₃₂₈, α₃₂₉, α₃₃₀, α₃₃₁, α₃₃₂, α₃₃₃, α₃₃₄, α₃₃₅, α₃₃₆, α₃₃₇, α₃₃₈, α₃₃₉, α₃₄₀, α₃₄₁, α₃₄₂, α₃₄₃, α₃₄₄, α₃₄₅, α₃₄₆, α₃₄₇, α₃₄₈, α₃₄₉, α₃₅₀, α₃₅₁, α₃₅₂, α₃₅₃, α₃₅₄, α₃₅₅, α₃₅₆, α₃₅₇, α₃₅₈, α₃₅₉, α₃₆₀, α₃₆₁, α₃₆₂, α₃₆₃, α₃₆₄, α₃₆₅, α₃₆₆, α₃₆₇, α₃₆₈, α₃₆₉, α₃₇₀, α₃₇₁, α₃₇₂, α₃₇₃, α₃₇₄, α₃₇₅, α₃₇₆, α₃₇₇, α₃₇₈, α₃₇₉, α₃₈₀, α₃₈₁, α₃₈₂, α₃₈₃, α₃₈₄, α₃₈₅, α₃₈₆, α₃₈₇, α₃₈₈, α₃₈₉, α₃₉₀, α₃₉₁, α₃₉₂, α₃₉₃,

The pursuit of this objective of restoring the cleft defect to the natural bone state has led to the surgical use of exogenous cytokines. The goal of the use of bone inductor cytokines is to induce the mesenchymal stem cells of the host to produce bone without the necessity of placement of a large amount of bone graft matrix. Mesenchymal stem cells occurring in growing children and adults have been shown to be capable of forming cellular elements of the skeletal-muscular system (i.e., muscles, ligaments, tendons, bone, fat, and connective tissue). The goal of producing a recon-

struction of osseous cleft defects by having the stem cells of the host being properly induced has been a very interesting one that has brought into the regeneration equation many aspects of research in the basic science areas of molecular biology, tissue engineering, and immunology. The identification of the appropriate cytokine to produce the desired result from stem cell morphogenesis has now led to a whole new scientific discipline involving the classification and the proper codification of cytokines and chemokines as shown in the Cytokine Periodic Table (Tables 27.2a and b).



Fig. 27.23. Intraoperative view of the patient shown in Fig. 27.22 after closure of the soft tissue of the nasal floor. Two sponges containing BMP-2 are placed, one on the palatal and one on the labial as shown here



Fig. 27.24. A view of the collagen sponges containing BMP from the labial aspect. The sponges are placed next to the bone surfaces posteriorly and anteriorly along the cleftal wall and against the soft tissue of the nasal floor which has been previously closed

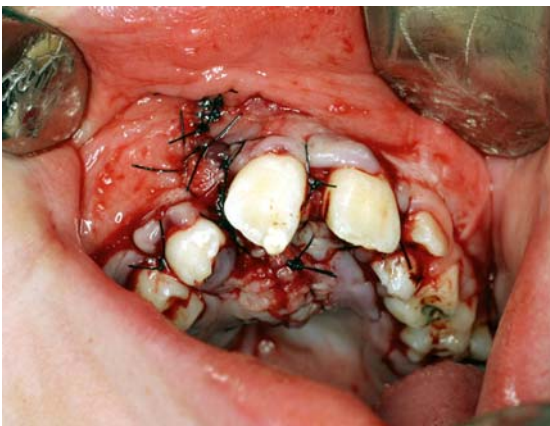


Fig. 27.25. The closure of the palatal and labial flaps



Fig. 27.26. A view of the closure of the palatal flaps which is very important in maintaining the cytokine in the area of maximal effectiveness against the bony surfaces of the cleft

27.10 Conclusions

With respect to the stability of the osseous segments in cleft palate cases, procedures involving bone grafting, bone manipulation, and orthopedic force application have changed remarkably during the last few decades. From the beginning of the 1900s, prosthodontic treatment was almost exclusively applied to “restore” the lost maxillary bone and reconstruct the osseous defect. After the WWII, bone grafting using large solid pieces of graft material were used for approximately two decades. These grafts were found to be unsatisfactory [6, 7]. In 1970, the application of osseous particulate autogenous grafts (PMCB) from the iliac crest was found to be remarkably successful in restoring the osseous defect [8–10].

It now appears that the next advancement in future treatment for the restoration of the osseous cleft defect will not involve actual bone grafting, but the use of cytokines to induce the mesenchymal stem cells of the patient to reconstruct the osseous defect.

In addition, the application of other genetic treatment modalities that have as their base the stimulation of the stem cells of the host are continuing to be developed and improved. Of these future genetic engineering treatment methods cell enhancement through tissue culturing, gene therapy, and more extensive use of exogenous cytokines to induce stem cells to form bone are very promising.

27.11 Summary

With appropriate early grafting of the cleft palate child and stimulation of the graft orthodontically and orthopedically, later orthognathic surgical procedures can be avoided and the incidence of the relapse can be greatly diminished. If appropriate treatment protocols are followed, relapse and the loss of control of the premaxilla, deficiencies in growth of the premaxilla, and other functional impairments can be avoided.

Additionally, the future appears to be very exciting in the area of developing improved methods of restoration and rehabilitation of the osseous cleft palate patient involving genetic engineering resulting in minimizing complications and long-term relapse.

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28 Secondary Bone Grafting of Alveolar Clefts

FRANK E. ABYHOLM

Cleft patients with a permanent osseous defect of the alveolar arch and maxilla will, even after the best surgical and orthodontic treatment, be left with the following deficiencies:

1. Limited prospects for orthodontic treatment. The osseous defect makes a nonprosthodontic dental rehabilitation impossible and necessitates a dental bridge to close the gap in the dental arch.
2. Instability of the maxillary segments, particularly of the premaxilla in bilateral clefts.
3. Oronasal fistulae or mucosal recesses that impede oral hygiene.
4. Insufficient support of the alar base contributing to the nasal asymmetry.

Secondary bone grafting of alveolar clefts combined with subsequent orthodontic treatment, designed to obtain a nonprosthodontic dental rehabilitation and also eliminate the other deficiencies attributable to the osseous defect, was started in Oslo in 1977. Our procedure is based mainly on the principles laid down by Boyne and Sands [1, 2]. Our clinical experience with more than 1302 cleft sites has confirmed the functional responsiveness of the grafted tissue to tooth migration and orthodontic movement of teeth.

28.1 Surgical Technique

The cleft area is widely exposed through incisions along the edges of the cleft. The incision on the vestibular side is made along the gingival border. Posteriorly, the incision is extended to the first permanent molar where it is angled up into the sulcus (Fig. 28.1). To provide sufficient mobility of this flap, which is going to cover the graft, it is necessary to cut

through the periosteum at the base of the flap. Anteriorly, the incision is extended along the gingival border to the center of the cleft side central incisor. Vertical incisions are made along the edges of the cleft. On the palatal side, mucoperiosteal flaps are raised along the edges of the cleft. A wide exposure of the cleft area is achieved with these incisions.

During the exposure of the cleft every effort is made to avoid traumatizing the thin bone lamellae that cover the dental roots adjacent to the cleft. The nasal floor is reconstructed, if necessary, and pushed upward. On the palatal side, the mucoperiosteal flaps are sutured together with everting mattress sutures. This leaves a well-defined cavity, whose walls are periosteum and denuded bone.

While the surgeon is exposing the cleft, an assistant has harvested cancellous bone from the anterior iliac crest through a small incision. A trap door of cortical bone is raised, hinged on the inner edge of the iliac crest. Chips of cancellous bone are removed by a sharp spoon, leaving the inner and outer cortex of the iliac bone intact. The donor cavity is filled with hemostatic felt (collagen). The lid of cortical bone is replaced and fastened with sutures.

The alveolar cleft is completely filled with cancellous bone chips. The alveolar crest must be formed up to the normal height and thickness. To improve nasal symmetry, sufficient chips must be placed under the alar base.

The lateral mucoperiosteal flap is advanced to cover the cleft and is sutured to the smaller medial flap and to the palatal flaps. In this way, only attached gingiva will cover the marginal area of the cleft site where the canine later will erupt.

In bilateral clefts, both sides are operated at the same time using the same operative technique.

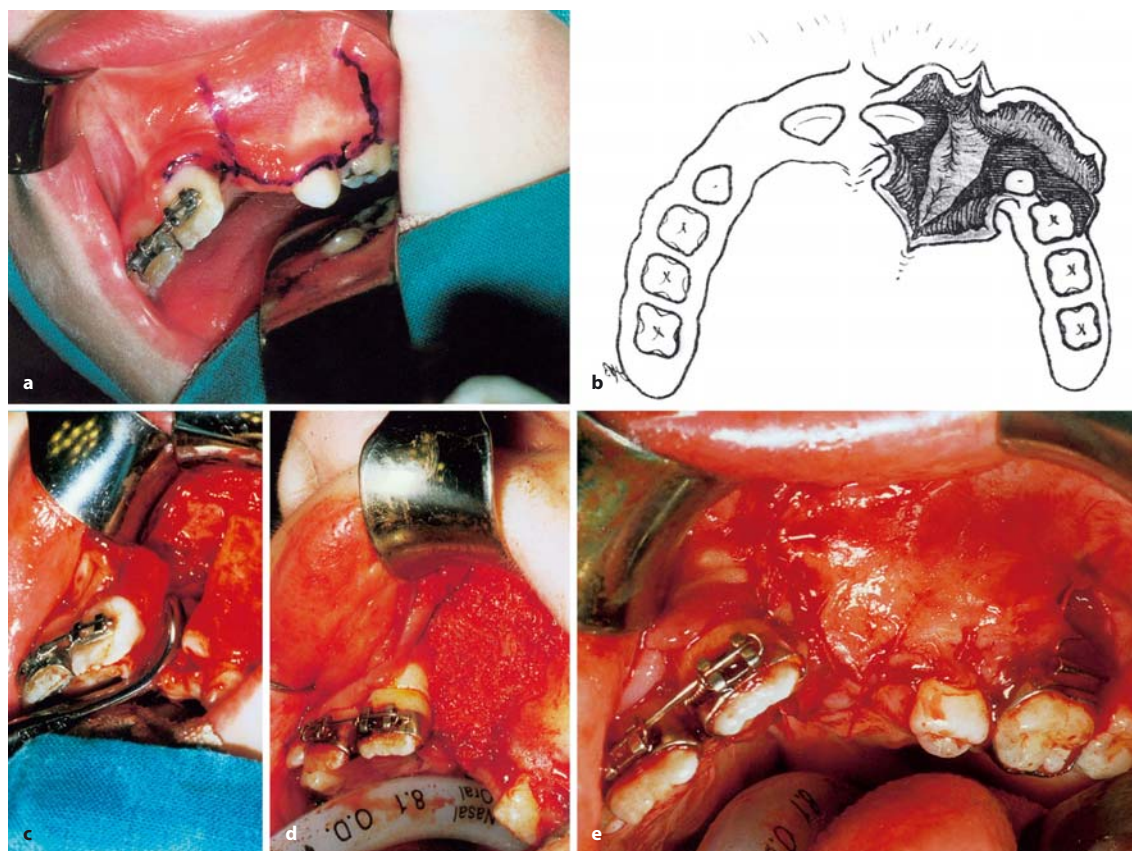


Fig. 28.1 a–e. Surgical technique: **a** Incision lines. **b** Schematic drawing of the raised flaps. **c** The alveolar cleft denuded. All soft tissue has been removed. **d** The alveolar cleft packed with can-

cellous chips. **e** The posterior flap transferred to cover the graft site. Only attached gingiva covers the cleft marginall

28.2 Orthodontic Management

28.2.1 Preparation for Bone Grafting

Orthodontic treatment is initiated in the early mixed dentition. Fixed appliance therapy alone has been used.

The maxillary incisors, which often erupt rotated, retroclined, and in anterior crossbite, are corrected for aesthetic reasons and to facilitate oral hygiene. Treatment generally lasts 3–4 months.

Approximately 25% of our patients with complete clefts of lip and palate have buccal crossbites attributable to segmental displacement.

Segmental repositioning is done just prior to the bone grafting procedure, and the orthodontic appliance is worn for 3 months postoperatively to retain the arch form. After this time, the bone seems able to retain the transverse dimension of the basal bone (any dentoalveolar relapse is corrected later). In patients

with bilateral clefts, a mobile premaxilla is stabilized with a heavy rectangular arch wire for 3 months postoperatively.

28.2.2 Permanent Dentition Management

When the cleft-side canine has erupted spontaneously, orthodontic treatment is resumed. The alignment of the permanent teeth follows different principles, depending on whether orthodontic or prosthodontic space closure is planned. If at all possible, we prefer orthodontic space closure because we consider bridgework in young adults to be undesirable for a number of reasons. In all patients with a missing lateral incisor in whom orthodontic space closure seems to be possible, every effort is made to move the posterior teeth forward. For these patients, we have found the Delaire-style protraction head gear to be useful. We regard this face mask as an adjunct to orthodon-

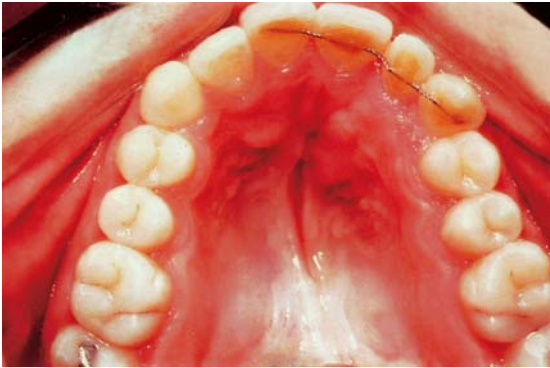


Fig. 28.2. When the orthodontic treatment is finished, a bonded palatal retainer is placed. This retainer is unobtrusive and does not interfere with oral hygiene

tics and not as a means of achieving clinically significant change in skeletal relationships.

In most patients, the treatment period in the permanent dentition lasts 2 years and is completed at the age of 15. A bonded palatal retainer extending to two teeth on either side of the cleft is placed. The retainer

is kept in place as long as the patient will accept it. Such retainers are unobtrusive and do not interfere with oral hygiene (Fig. 28.2).

28.3 Optimal Age for Secondary Bone Grafting

In the discussion of this subject two factors are of particular importance: (a) the curve of growth of the various growth sites in the maxillary complex, and (b) the clinical goal of the bone grafting.

Possible interference with postnatal maxillary growth must be considered in identifying an optimum or ideal age for secondary bone grafting. Sagittal and transverse growth of the anterior maxilla has virtually ceased by the age of 8–9 years [3, 4]. The vertical growth of the maxilla occurs mainly as deposition of additional alveolar bone at the alveolar crest [5]. The continuous eruption of teeth is thought to be the agent that stimulates the formation of alveolar bone. With a viable donor tissue such as autologous cancellous chips, the graft is rapidly transformed into functional alveolar bone responding physiologically to erupting teeth (Fig. 28.3). By spontaneous or ortho-

Fig. 28.3 a, b. The grafted bone responds physiologically to the erupting canine: **a** Alveolar cleft prior to bone grafting. **b** The canine erupting normally through the grafted bone



dontically guided eruption of the canine, the capacity of erupting teeth to generate alveolar bone can be utilized to maintain the general growth in maxillary height.

Cephalometric studies of the Oslo cases have shown that bone grafting of the alveolar cleft following the principles stated earlier and performed between the ages of 8–11 years had no adverse effect on anteroposterior or vertical maxillary growth [6].

Some authors advocate bone grafting at the age of 5–6 years in order to give any lateral incisor an opportunity to migrate into and erupt through the bone graft. This is a strong argument from a dental point of view. However, further studies are necessary to prove that bone grafting at this age does not interfere with maxillary growth.

A primary goal of secondary bone grafting is achieving orthodontic closure of the cleft space. Orthodontic space closure has proven to be easier when bone grafting is performed prior to the eruption of the cleft-side canine. In our studies, in the group in which bone grafting was performed before the full eruption of the canine, orthodontic closure of the gap was possible in 93% of the cases (335 out of 360). In the group in which the bone graft was performed in the permanent dentition the success rate was 72% (107 out of 149) [7–9].

An evaluation of the height of the interalveolar bony septum also showed a difference between the two groups with a better height of the interalveolar septum in patients who had the bone grafting performed before the full eruption of the cleft-side canine [7–9].

28.3.1 Secondary Bone Grafting

After 25 years of bone grafting in the transitional dentition we have analyzed 1070 patients, of whom 232 had bilateral clefts, making the total number of grafted cleft sites 1302.

Of these, the teeth in the cleft region were in the final position in 992 cleft sites and thus the interdental septum could be evaluated. Good bony interdental septum height (3/4 or more of normal septum height) was found in 96% when the operation was performed before the eruption of the canine, in 85% when the operation was performed after the eruption of the cleft-side canine. Failure rate: no bone formation in 1.2%. A complete dental rehabilitation without prosthodontics was achieved in 93% of the patients when the bone grafting was performed before the eruption of the cleft-side canine.

In order to investigate the fate of the grafted bone, we have examined 18 consecutive patients with uni-

lateral complete CLP with 3D CT scan 20 years after bone grafting [10]. We found good bony support of the teeth adjacent to the cleft in all cases, but there was less bone in the cleft area than on the normal side. The apertura piriformis was lower on the cleft side in all cases, and there was hypoplasia of the maxilla on the cleft side.

28.4 The Grafting Tissue of Choice

When autogenous cancellous bone is transplanted under optimal conditions, osteogenic cells in the graft will survive, and new bone formation will start within a matter of days. Cancellous grafts subjected to minimal traumatic surgery undergo a rapid revascularization which is important for the long-term result [11]. Fresh autologous cancellous bone transforms very rapidly into alveolar bone, which responds normally to tooth migration and orthodontic movement of teeth.

A cortical bone graft undergoes a slow transformation. The early establishment of nutrition to the cortical bone cells requires restoration of flow through existing vessels or canaliculi and ingrowth of capillaries. This is a very slow process, and the cortical bone will usually die and be replaced by invasion of bone cells originating from the recipient site. This is a fundamental difference between cortical and cancellous bone grafts in terms of both cell survival and vascularization. The slow transformation of a cortical graft makes the reestablishment of the tooth-bearing function of the alveolar process unfeasible.

28.4.1 Donor Sites

For autotransplantation of bone in cleft patients several donor sites have been exploited: rib, tibia, skull, chin, and iliac crest. All of these sources have been used successfully. We have, however, found the iliac crest to be the most suitable donor site.

28.4.2 Complications

Serious complications are rare. Failures (i.e., no continuous bone bridge across the cleft) are probably due to poor surgical technique or infection. External cervical root resorption can occur if the root cementum is mechanically injured during the bone grafting procedure. This can be avoided by delicate surgical technique and by performing the operation at an age when the cervical region of the canine is still covered with bone [8].

28.5 Important Surgical Details

To achieve optimal results, we have found some details to be of great importance.

28.5.1 Flap Design

It is important that the mucoperiosteal flaps are designed in such a way that only attached gingiva will cover the marginal part of the graft. It is our clinical observation that a completely normal periodontium seems to occur mainly in cases where the flap design has ensured an orthotopic transfer of attached gingiva to cover the marginal part of the graft.

28.5.2 Good Exposure

The cleft must be widely exposed so that all scar tissue in the cleft area can be removed. The nasal floor has to be reconstructed if a fistula is present. It is important to elevate the nasal floor to achieve a good height of the alveolar crest so that the canine can be brought into a vertical position in the cleft area.

28.5.3 Cancellous Bone Only

Only autologous cancellous bone creates bone that responds normally to eruption and orthodontic movement of teeth.

28.6 Conclusion

Autologous cancellous bone grafted to the alveolar cleft between the ages of 8–11 years creates alveolar bone which responds normally to eruption and orthodontic movement of teeth. A full dental rehabilitation is possible in the vast majority of cases when bone grafting is performed prior to the cleft-side canine eruption, rendering bridgework unnecessary in cleft patients (Fig. 28.4).

Bone grafting performed between the ages of 8–11 years does not restrict maxillary growth.

This combined surgical/orthodontic treatment plan requires close cooperation between plastic surgeon and orthodontist. Fringe benefits include: closure of oronasal fistulae and elimination of mucosal recesses, stabilization of maxillary segments, and improvement of nasal symmetry.

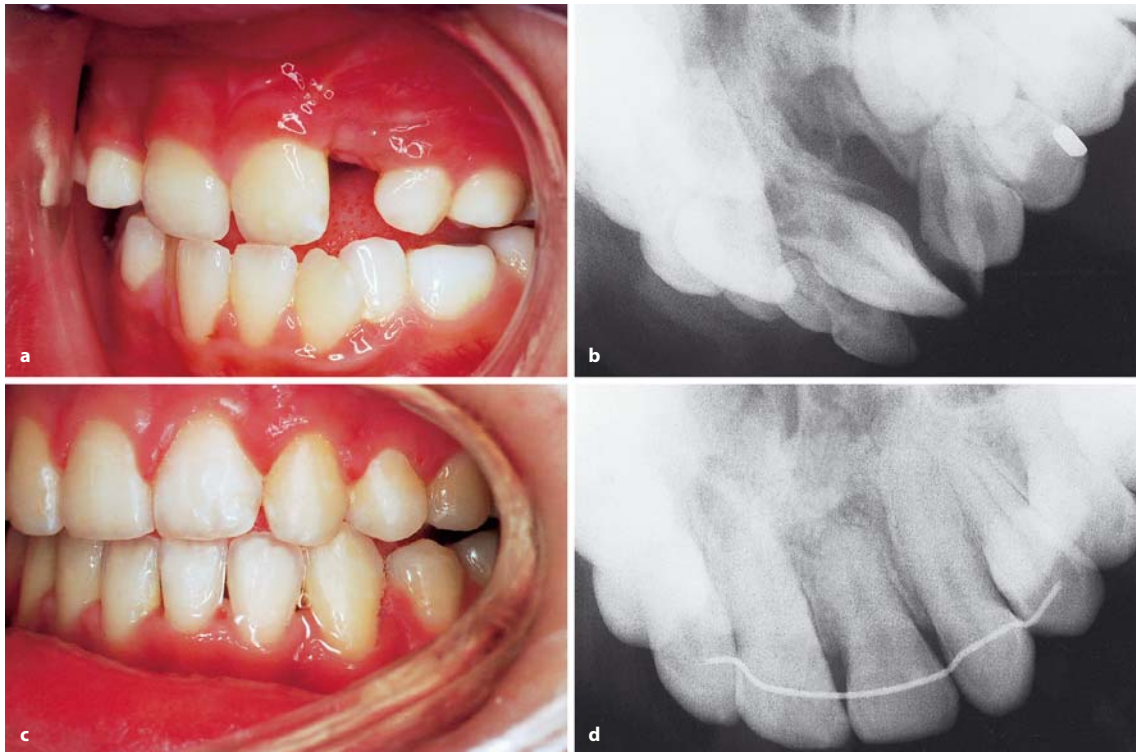


Fig. 28.4 a–d. Unilateral complete cleft bone grafted prior to the eruption of the canine: **a** The dental arch before the bone grafting. **b** Radiograph of the cleft prior to bone grafting.

c, d Dental arch and radiograph after the completion of the orthodontic treatment

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29 Speech Implication of Orthognathic Intervention

DONNA RUSSELL FOX

Speech production is much like looking through a window to the scene beyond, an invisible means of conveying an articulate, intelligible message in a seemingly effortless and inconspicuous manner. This is the standard normal speech, the goal which clinicians who deal with craniofacial anomalies must keep in mind. The production of normal speech is the ultimate test of the effects of reconstructive and rehabilitative efforts.

This chapter will focus on three topics: the role of the speech-language pathologist on the craniofacial team; the effects on speech when the vocal tract is altered by malformation, trauma, or surgery; and some considerations for orthognathic surgeons as they prepare patients for surgery.

Craniofacial anomalies are complex structural and functional abnormalities. Because of their complexity, the skills of many specialists are required in patient management. Each specialist has his or her unique goals based on special skills and training. Plastic or oral surgeons seek to optimize aesthetic appearance; dental specialists seek to improve appearance and obtain function in biting and chewing; and the speech-language pathologist oversees the patient's language acquisition and speech production.

29.1 Role of Speech Language Pathologist

The development of normal speech is influenced by the child's innate language ability, intellectual capacity, hearing acuity, articulation competence, velopharyngeal adequacy, oral motor coordination, and vocal tract configuration. Speech development is optimal when the underlying vocal tract is anatomically and physiologically intact and accompanied by the intact kinesthetic and auditory feedback systems. Ideally, these feedback systems are established early in life resulting in articulate, intelligible speech [1]. Devia-

tions, either congenital or acquired, can affect speech output. When there is dysfunction in one of the speech systems, there is rapid compensation in the function of other systems, due to their interdependence and integration. When compensation cannot overcome the dysfunction, speech and language disorders become apparent.

The vocal tract consists of the lungs and thorax as the energy source; the larynx for phonation production; the pharynx, nose, and mouth for resonance; and the tongue, teeth, lips, jaw, and palate for articulation. One of the contributions of the speech-language pathologist to the cleft palate-craniofacial team is to interpret observations and testing of the vocal tract so that strategies may be planned in conjunction with all members of the team to maximize speech outcome and complement the aesthetic outcomes. The speech language pathologist documents the patient's speech and language status both initially and over the course of intervention; however, the vocal tract configuration is of primary importance to orthognathic surgeons, and it is these structures that merit their immediate concern and are the primary focus of this chapter.

29.2 Effects of Altering the Vocal Tract on Speech

The second topic considered in this chapter deals with the effects on speech of orthognathic surgery on the vocal tract. Examples can be taken from the literature regarding the changes surgery can produce on the lips, tongue, teeth, and palate, but most particularly on the function of the velopharyngeal port.

The first surgical procedure to affect the lip is lip adhesion or primary lip repair in the cleft lip patient. We know that this closure tends to round and retroposition the arches in a position closer to normal, and thus provides the possibility of normal lip closure and pressure. Profit and Phillips [2] noted that "Ortho-

gnathic surgery has the potential to affect the lip position and thereby alter this equilibrium because it produces both lip movement in response to movement of the teeth and jaws and independent lip movement caused by contraction of soft-tissue incisions or a change in muscle tonicity." Superior repositioning of the maxilla, advancement of the mandible by ramus osteotomy, and advancement of the maxilla tend to stretch the lip and increase tension on the lips. However, Profit and Phillips [2] did not find a significant increase in lip pressure when the jaws and teeth were moved in a way that stretched these soft tissues. In contrast, they reported that, although the relaxation of soft tissues that accompanies superior repositioning of the maxilla could be seen clearly in lower resting lip and cheek pressures, it was not observed during speech and swallowing, which they interpreted to indicate that the degree of muscle contraction necessary to obtain an appropriate lip seal is monitored and maintained. A more recent article by Ewing and Ross [3] documented soft tissue changes in LeFort surgery. One of their important conclusions was that, even with measurements of the soft tissue ratios between maxillary advancement and soft tissue response, these were not all dependable. This may be the result of, in this case, the kinesthetic feedback system, which adjusts and maintains postures. For an example of this concept, see the pre- and postsurgery photographs in Fig. 29.1 and note the lip positions.

The sounds most affected by lip positions are /p/, /b/, /m/, /f/, and /v/; therefore, these sounds are productions to particularly note in lip surgery of the older patient. One would predict that production of these sounds would improve as the lips are brought into approximation. If, however, the sounds were defective before surgery, the same production patterns may persist after surgery. The possibility for correct pro-

ductions, however, is increased with improvement in structure.

The subject of the tongue has long held the interest of surgeons as well as speech-language pathologists, and the literature is far too extensive to review here. Suffice it to say that ablative surgery appears to have the greatest effect, with disability increasing with larger amounts of tongue resection, and total glossectomy affecting speech intelligibility to the greatest degree [4]. The mobility of the tongue appears to be the key to oral cavity function (speech, swallowing, chewing, etc.) [5]. Argamaso [6] followed by LeBlanc and Golding-Kushner [7] reported on effects of glossopexy in 17 patients with Robin sequence between 2 and 16 weeks whose tongue release and palate repair ranged from 9 to 15 months. Sound productions were recorded pre- and postsurgery with normal speakers used as controls. Most of the patients with Robin sequence showed no difference in sound production within 12 months of glossopexy detachment. This, again, is an example of the ability of the oral mechanism to make adjustments for changes in the oral environment. The tongue is the most important organ for the correct place and manner of sound production. Thus, speech can be intelligible even though the voicing and resonance characteristics are abnormal. An example of this is an individual with cleft palate whose speech is easy to understand, but whose voice quality is noticeably nasal. Again, compensatory feedback systems can take over for other systems only within certain limits.

The mechanism that captures the avid interest of everyone who diagnoses and treats cleft palate patients is the velopharyngeal port (VP). The question for the orthognathic surgeon is: if the maxilla and/or mandible are moved, will the VP port change and thus affect speech, most particularly its resonance charac-

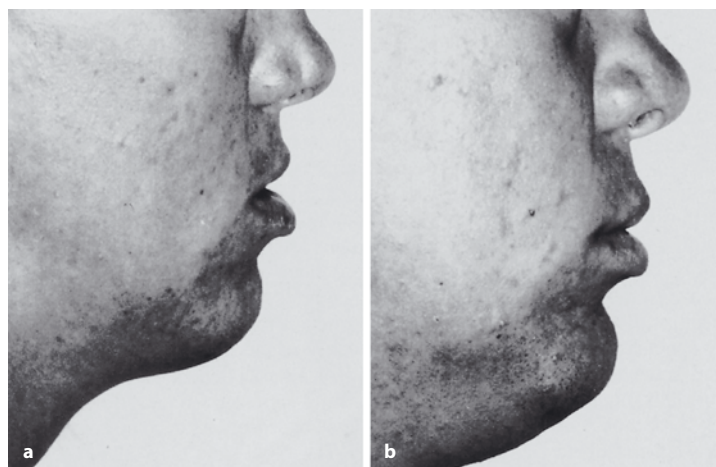


Fig. 29.1. Maintenance of lip postures following surgery. **a** Presurgery. **b** 1½ years postsurgery

teristics? Some researchers have questioned whether the VP mechanisms are compromised in cleft palate [8–13]. Kummer et al. [12] for example, indicated that the seven cleft patients in their study did not show increased risk for postoperative hypernasality, although they did show deterioration and change in the velopharyngeal port, which the authors interpreted to be of no clinical significance. In a commentary on the Kummer et al. article, published at the same time, Witzel [14] disagreed with this interpretation.

Articulation, however, has been reported to improve in most, but not all, cases where surgical improvement of occlusal defects has been studied. Both Witzel [15] and Vallino [13], as well as others, continue to support the concept that orthognathic surgery for malocclusions has deleterious effects on resonance, but support the concept that this surgery leads to improvement in articulation. In noncleft patients, most authors have indicated that there is little risk of decompensation of the velopharyngeal port.

In a 1964 article, Schuchardt [16] questioned the effect of moving the maxilla on the velopharyngeal port, and he was followed by Converse et al. [17], Schwarz and Gruner [18], and Epker and Wolford [19]. In their study of osteotomies of the middle third of the face, Epker and Wolford [19] indicated that the velopharyngeal portal was at risk, but unfortunately their data cited did not separate cleft from noncleft patients. Other studies have indicated some deterioration of the velopharyngeal closure [9, 10, 15, 18, 20–22]. Witzel and Monroe [20] clearly documented that the forward shift of the maxilla produced velopharyngeal inadequacy and nasal resonance. In addition to her commentary on the 1989 article of Kummer et al. [12] in which Witzel [14] disagreed with the finding of “no clinical significance,” she provided additional data on patients treated at the Hospital for Sick Children. In a sample of 41 patients without cleft palate who had LeFort 1 maxillary advancement and 50 patients with repaired cleft palate or cleft lip and palate, the patients without cleft palate had a very low risk for deterioration of velopharyngeal function, as did patients with a repaired cleft palate who had adequate VP function before surgery. The 11 of 15 cleft patients in Witzel’s “borderline” preoperative ratings who acquired postoperative VPI, along with the patients who had inadequate VPI pre- and postoperatively merit our concern. This concern is given added strength by two recent articles. Watzke et al. [23] reported that 5 of 24 cleft patients who underwent maxillary advancement experienced deteriorated velopharyngeal function. Of the 5, 4 have had velopharyngeal function prior to surgery. Okazaki [24] reported that 8 of 10 cleft palate patients in this study showed increased hypernasality after surgery.

More recently the use of maxillary distraction osteogenesis has become popular as a method of changing the maxillary or mandibular processes. Various methods have been reported in the literature and changes in technique appear with some frequency. However, only a very few studies have emerged to test the effect of these techniques on the vocal tract and speech output. Ko et al. [25] indicated in their study of 22 patients that speech evaluation was an important aspect in planning any maxillary distraction because distraction can influence the velopharyngeal closure and thus the resonance effect in speech. Guyette et al. [26] studied distraction on the variables of resonance, articulation, and velopharyngeal function, and concluded that the distraction and LeFort procedures carried similar risk for these speech changes. The most recent study is by Harada et al. [27]. In a series of six patients, he reports that a distraction of less than 15 mm did not compromise velopharyngeal function. Only 1 of the 6 had an increase in hypernasality, and this patient was hypernasal prior to surgery. Guyette [28] and Harada [27] both suggest that velopharyngeal incompetence may be related to the amount of distraction, particularly if the amount is close to 14–15 mm.

The 2002 study by Sell et al. [29] provides an excellent bibliography on techniques for maxillary advancement, as well as including their use of transpalatal advancement without creating velopharyngeal dysfunction (nasal resonance, nasal friction, or misarticulation). However in studies do not differentiate between resonance, articulation, and velopharyngeal function, but lump all speech attributes together, the final outcome is difficult to assess and the comparison of studies is made difficult.

In summary, there are numerous examples demonstrating the effects of orthognathic surgery on speech, but as we have become more sophisticated in both our measurements and our perceptions, research underscores the need to evaluate patients carefully prior to intervention. When the bones and muscle of the oral cavity are changed, changes occur in velopharyngeal contact, thickness, and height as well as an increase in gap between velum and pharyngeal wall. Other studies [12] have reported velar lengthening. What then can we use to help predict the possible effects of surgery?

At least five factors can aid us in sorting out patients who are at high risk for deleterious effects of orthognathic surgery. The first factor can be found in the presurgical history. If there is a history of speech problems in the family and/or in the patient, presurgical documentation of the exact nature and status of the patient’s language, resonance, articulation, and intelligibility is necessary. A second factor is the quality of the VP contact. The velopharyngeal

port must be studied and, to some extent, measured by fluoroscopy and/or endoscopy. We know that patients with good preoperative velopharyngeal contact are at less risk for postsurgical problems than are patients with borderline or poor contact. A third factor is that the projected movement of the mandible and maxilla needs to be estimated presurgically and probable changes in its postsurgical relations to the other oral structures evaluated. Precise postsurgical relationships are impossible to predict accurately, but estimates can be used to counsel patients on potential effects on speech. Fourth, the presence or absence of a pharyngeal flap will probably affect the final outcome of the surgery. The presence of a flap tends to indicate more risk to the velopharyngeal port. Finally, the presence of other factors, such as occult or submucous cleft or the presence of a syndrome, are indicative of higher risk for postsurgical resonance deterioration.

Two anecdotes with very different scenarios provide examples of the possible risk factors in patients. Case 1 is a boy 14 years of age who has a cleft in the lip and palate, but has had good surgical repair and speech intervention and has worked hard on having what is now normal speech. The aesthetic result of the deformity, however, is less than acceptable, and he is in need of a LeFort I procedure. However, after an explanation of the risks because he has borderline velopharyngeal competence, he decides that, having worked so hard to achieve normal speech, he will not have the surgery. A year goes by filled with rejection by classmates (particularly girls), and he again talks to the surgeon who feels that surgery can be done without too much risk to the velopharyngeal port. In addition, he is told that the possible resulting VIP could be reduced with a flap or an implant. The surgery is accomplished and indeed the patient looks considerably better and his speech remains the same. In this case, the outcome was an excellent one.

The second case is a young woman who wishes to improve her appearance. She is a homemaker with two children and not employed. In this case, a LeFort procedure was performed along with a genioplasty. Following surgery, the patient complained that her voice quality had changed. She had been a professional singer and had taken off several years to raise her children, although she had continued to volunteer for the church choir, serving as the musical director. She now feels that the quality of her voice has changed, she has reduced sensation in her lower lip, and at times experiences some drooling. At the time of her evaluation by the speech-language pathologist she was about 1 year postsurgery. The patient was not happy with the quality of her voice, and she had not been warned of the possible effect the surgery might have on her voice. She is presently suing the surgeon. Here

the outcome was an unhappy one. A better preoperative history and good speech and voice evaluation might have prevented this unfortunate outcome.

29.3 Preoperative Considerations for Orthognathic Surgeons

The final topic deals with considerations for orthognathic surgeons as they prepare patients for surgery. A careful preoperative history should be taken, particularly regarding patients' previous surgery, speech intervention, and professional aspirations. All patients should undergo at least a screening procedure of speech and language, as well as resonance prior to surgery. In patients for whom the risk factors outlined above are high, a detailed objective and subjective evaluation of the speech mechanism, both pre- and postoperatively, should be undertaken. It should include some documentation regarding the function of the entire vocal tract, but most particularly the use of videofluoroscopy and or endoscopy to study velopharyngeal function and to document and assess the quality of the velopharyngeal closure.

Standard and thorough explanation of possible changes in articulation, resonance, quality, and intelligibility should be routinely given prior to surgical intervention being considered. We must expand our concern, not only for the cleft patients with whom we are involved who present with high risk factors, but also to the other part of the aesthetic scale. We must be mindful of the effects of the surgical procedures on professional speech and voice users for whom even slight changes in resonance may have devastating effects on career and salary.

The statements we can make based on information in the literature is that patients *probably* can be improved in the areas of lip posture, occlusion, articulation, and airway with orthognathic surgery. Secondly, VPI may occur in some patients with borderline VP closure and result in hypernasality. The resulting hypernasal resonance problem may or may not be improved with speech intervention or secondary procedures such as pharyngoplasty, implantation, etc. Third, patients with a small presurgical gap may find that the gap has increased postsurgically. Fourth, patients with VPI preoperatively will continue to have VPI postoperatively.

In light of the foregoing information, it is evident that considerably more research is needed in all of these areas, but most particularly in developing definitive measures of the lip, tongue, and VP movement and function following surgery. Susami et al. [30] have reported some improved measures of lip elasticity in repaired cleft lip patients, and the photodetection method reported by Karnell and Seaver [31] is prom-

ising for assessing VP function. Predictive measures used presurgically to investigate risk also deserve further investigation. Sandor et al. [32] reported on the validity of using nasendoscopy as a predictive measure, and Ewing and Ross [3] have documented the difficulty in attempting to predict soft-tissue response to surgery. Shprintzen [33] provides some meaningful considerations concerning pharyngeal flaps and reiterates the lack of scientific data regarding the stability of prediction for outcome. We need careful and well-documented speech results before and after surgery, including articulation, resonance, vocal quality, and intelligibility. Finally, what is the effect of relapse? That is a question that deserves an entire book. Perhaps in the near future we can look forward to a more positive statement about the effects of surgery on VP function than is possible at the present time.

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30 Diagnostic Procedures and Instruments Used in the Assessment and Treatment of Speech

SAMUEL BERKOWITZ

30.1 Articulation Tests

An articulation test is a paper and pencil test used by speech-language pathologists to systematically evaluate the formation and production of the sounds of speech in different contexts in words and sentences. It is used to record the sounds that are produced correctly as well as the errors in speech including omissions, distortions, and substitutions of normal or compensatory sound errors. Systematic evaluation of the articulation of speech is helpful in ensuring complete and consistent analysis of the problems so that effective and efficient treatment can be planned.

Many different types of tests are available, but the most useful tests for patients with cleft palate are those that assess both place and manner of production of speech sounds in the vocal tract. Individuals with cleft palate often produce sounds in a more anterior or more posterior (this is a more frequent error) location in the vocal tract than normal speakers. Careful notation of the types of errors and the probable causative factors sets the stage for determining the method and duration of treatment. These tests also provide a baseline measure or description of speech articulation against which progress, improvement, developmental changes, and treatment outcome can be evaluated. The examiner scores articulation of individual sounds or sound units by listening to and observing the production of these sounds. In some cases, traditional articulation testing alone cannot completely describe the manner or place of the error sounds and visualization techniques such as multi-view videofluoroscopy and nasopharyngoscopy are useful in completing the description and diagnosis of the error(s).

30.2 Rating Scales of Speech Intelligibility and Acceptability

Rating scales are often used by speech-language pathologists and other members of the cleft palate team to score the overall severity of an individual's communication impairment in several categories. Ratings of intelligibility describe how well an individual's speech can be understood by others, whereas ratings of acceptability describe the pleasingness of the sound and appearance of speech. These scales usually consist of 5 or 7 points, with 1 being normal and 5 or 7 indicating unintelligible or unacceptable speech. Effective use of these scales is related to rater reliability, and raters must establish and re-establish their reliability on a regular basis when using these scales for clinical or research purposes. These scales are useful as overall measures of severity and treatment outcome; however, they are descriptive in nature and are not useful in isolating causative factors.

30.3 Cephalometrics

Orthodontists and specialists in facial growth have developed the technique of cephalometrics. This is a still sagittal x-ray taken with the patient stabilized in a headholder which positions the head relative to a cranial landmark called the Frankfort horizontal plane. The head is secured by earposts, and the subject's midline is positioned at a constant distance from the x-ray source. The standard position and photon source relationship to the subject provide a means of comparing the measurements from the resulting x-rays. It should be understood that x-ray films provide an image that is a summation of the tissue through which the x-ray beam has passed. If a person had soft palatal closure on one side but not on the other, the midsagittal view would show velopharynx-

geal closure because the beam would pass through the tissue on the closed side, and the resulting image would not yield information about the opening. The velopharyngeal valve is three dimensional, and its attributes cannot all be captured in the midsagittal view alone. This is a shortcoming of cephalometrics when used for speech purposes. Another problem with the technique is that it is capable of filming only a single point in speech production. Connected discourse cannot be studied by this means, nor is it possible to specify precisely what will be filmed in the course of production of a single sound.

Of particular interest to speech physiologists are the cephalometric studies that have defined structures and growth patterns of pharynx, velum, and lymphatic tissue [1–4]. Compositely, these cephalometric studies have provided objective data pertaining to normal skeletal and soft tissue structures and their postural relationships.

This information also has been stated within the complex and important reference of a specified stage of growth and development. The diagnostic value of such normative data becomes obvious in considering the oral examinations speech-language pathologists routinely perform to evaluate speech structures. In such examinations, skeletal, dental, and soft tissue components of the speech apparatus are evaluated.

In addition to the normative data provided, cephalometric analyses have also been undertaken to evaluate skeletal characteristics and growth disturbances in pathologic conditions. Especially valuable for speech-language pathologists are the studies which have defined the intraoral, intranasal, and pharyngeal architecture of individuals with clefts of the lip and palate [5–9]. Speech-language pathologists have learned a great deal about morphologic variation in cleft palate patients, whose total rehabilitation, of necessity, includes multiprofessional concerns, from these cephalometric studies.

The fact that cephalometric head plates can be reliably compared has made them attractive to speech-language pathologists intent on defining physiological differences that result in defective speech production.

There is probably universal agreement that a midsagittal cephalometric view, although useful, yields incomplete information about soft palate and lateral pharyngeal valving. It provides little insight into the location, configuration, or movement of structures off the midsagittal plane. In particular, it offers no information about movement of the lateral pharyngeal walls.

30.4 Cine- and Videofluoroscopy [10]

Cinefluoroscopy (x-rays recorded on motion picture film) and videofluoroscopy (x-rays recorded on videotape) with simultaneous voice recordings are useful procedures in the evaluation of individuals with cleft palate. The key to development of motion x-rays was the advent of image intensification, which permitted greater contrast with reduced x-ray dosages. Video recording involves lower radiation dosage than does cinefluoroscopy. Videofluoroscopic recording can be done with less radiation than is required for a single still x-ray of the head.

To extract more information from videofluoroscopy, Skolnick [11] introduced multiview videofluoroscopy, a technique that adds a base view to the traditional lateral and frontal projections.

30.5 Multiview Videofluoroscopy

This fluoroscopic x-ray technique allows visualization of the velopharyngeal port, or valve, during speech in several different planes [11, 12]. This is important because the mechanism of velopharyngeal closure is three-dimensional and may involve movement not only of the soft palate, but also of the lateral and posterior pharyngeal walls. Descriptions of the technique include the lateral, base, Towne's, frontal, Waters', and oblique views. Not all views are required for a complete examination. The view selected depends on the information needed, the anatomy of the skull, and the anatomy and function of the vocal tract. In most cases, a complete examination consists of three views, including the lateral view; the base, or Towne's view; and the frontal, Waters', or oblique view. The examinations are usually recorded and maintained on videotape with audio recording for purposes of interpretation and comparison. The procedure is conducted and interpreted jointly by a radiologist and speech-language pathologist. This contributes to a valid study which includes an adequate speech sample, appropriate visualization of the velopharyngeal valve, and interpretation of the findings which are consistent with the patient's speech. Radiation dosage is kept to a minimum by using videofluoroscopy, as opposed to cinefluoroscopy; coning the x-ray beam to the smallest area, ensuring that the equipment is emitting the minimum radiation necessary to produce the image; and using lead sheets to shield the patient except for the area of interests.

30.5.1 Technique

To enhance the velopharyngeal area, high density barium is instilled into each nostril using a syringe with a plastic tip. This results in barium coverage of the soft palate, lateral and posterior pharyngeal walls, and posterior aspect of the tongue. A standard speech sample is used during each view. The lateral view is obtained first, with the patient sitting upright. In this view, judgments can be made about the length and thickness of the soft palate, the depth of the pharynx, and the size and location of adenoid and tonsillar tissue. The anterior-posterior excursion of the soft palate and, in some cases, the anterior movement of the pharynx are observed and judgments of velopharyngeal contact during speech are made. This view alone is not sufficient to determine velopharyngeal closure because it does not confirm velopharyngeal contact along its width.

The lateral view permits excellent visualization of the function of the tongue during speech and is helpful in the differential diagnosis of compensatory articulation errors. In some cases, the tongue is observed to elevate the soft palate in an attempt to effect velopharyngeal closure, particularly for the velar stop sounds /k/ and /g/. Abnormal posterior movements of the epiglottis and elevation of the larynx during speech are also apparent in this view.

The frontal view is usually performed next to determine the degree and location of mesial movements of the lateral pharyngeal walls. Lateral pharyngeal wall movements may occur at a specific location in the vocal tract or along an extensive area. This information is thought to be useful in planning the approximate width of a pharyngeal flap or designing a prosthetic speech appliance in cases of velopharyngeal insufficiency. To obtain this view, the patient is seated upright facing the image intensifier with his or her head positioned in the Frankfurt horizontal plane. Complete coating of the nasopharynx with barium is essential to gather information for adequate interpretation. The standard speech sample is used.

In cases where the overlying bony structures obscure the lateral pharyngeal walls and their movements, the Waters' view is a helpful alternative. In this view, the head is tilted upward approximately 45° from the Frankfurt horizontal plane so that the bony structures do not impede the view of the lateral walls. When asymmetric movement of the lateral pharyngeal walls is observed, Shprintzen et al. [13] recommend rotation of the head to the right and left along the x axis to more clearly identify the extent of movement.

The third view to be performed is either the base or Towne's view. These views outline the shape of the velopharyngeal valve; the pattern, symmetry, and

consistency of velopharyngeal valving movements; and the size and location of velopharyngeal gaps during speech. The Towne's view is the more useful of the two views when the soft palate approximates adenoid tissue during valving creating an oblique axis of velopharyngeal closure or when patients have large tonsils or posterior compensatory tongue movements. These situations interfere with adequate visualization and interpretation of velopharyngeal movements in the base view.

To obtain the base view, the patient lies prone on the x-ray table in a sphinx-like position with the head hyperextended. To obtain the Towne's view with overtable tube fluoroscopy equipment, the patient is seated upright with his or her head in the horizontal plane. The camera is then rotated in relation to the face until the velopharyngeal valve is visualized. This view is similar to that used in nasopharyngoscopy. The standard speech sample is used for both the base and Towne's views.

Multiview videofluoroscopy may be reliably performed in patients as young as 3 or 4 years old. Because this test involves radiation, it should be used judiciously. It is most reliable when the child is mature enough to cooperate for the examination and has sufficient speech development to allow visualization of the valve throughout all classes of speech sounds. The test should be used when the clinical speech assessment suggests velopharyngeal inadequacy, particularly when surgical or prosthetic management is being considered.

30.6 Ultrasound

Ultrasound is a device that has been employed in the evaluation of velopharyngeal function, particularly movements of the lateral pharyngeal walls. It is not suitable for displaying motion of the velum because of problems in transmitting ultrasound through bone overlying the palate [14].

30.7 Video Nasopharyngoscopy

This technique involves inserting a flexible fiberoptic tube into the nose to obtain a direct superior view of the velopharyngeal valve and vocal tract during speech. In the hands of an experienced and patient examiner, the test provides information about the anatomy and function of the velopharyngeal valve during speech; the relative size, location, and consistency of velopharyngeal gaps; the function of the posterior aspect of the tongue during speech, which is helpful in the differential diagnosis of compensatory articulation errors; and the anatomy and function of the

laryngeal structures. It is also useful in the identification of pulsations in the pharynx which may be indicative of abnormally placed carotid arteries that might preclude pharyngeal flap or sphincter pharyngoplasty surgery. Although the latter finding is rare, it occurs most often in patients with velocardiofacial syndrome. This technique is useful not only for pre-treatment diagnostic assessment of velopharyngeal function but also to evaluate the outcome of surgical, prosthetic, and/or speech therapy treatment. It may also be used for biofeedback therapy to enhance velopharyngeal movements during speech and correct some compensatory articulation errors. For ease of examination in young children, a flexible nasopharyngoscope with a distal tip diameter between 2 and 3.7 mm is recommended.

McWilliams et al. [15] note that lateral wall movement is not reliably assessed by either measurements or judgments of nasoendoscopic images. A major reason for using endoscopic procedure is to learn about the contribution of the lateral pharyngeal walls to velopharyngeal function. Some investigators have assumed symmetry of lateral wall movement in making their judgments, but this assumption is questionable. Further data relative to the realizability and validity of endoscopic measurements are needed. In the meantime, the technique is quite commonly used clinically and has much to offer in the hands of trained examiners.

30.7.1 Technique

A topical anesthetic such as 3% lidocaine or 2% tetracaine hydrochloride mixed in equal parts with .5% phenylephrine is applied through one side of the patient's nasal cavity to allow comfortable insertion of the scope and enhance cooperation. Some patients are able to tolerate the procedure without anesthesia, particularly when a smaller scope is used. The nasopharyngoscope is inserted through the middle meatus of the nasal passage to first visualize the velopharyngeal area to document anatomy and function and scan the pharynx for abnormal pulsations. The scope is then passed down into the vocal tract to document the anatomy and function of the tongue and larynx. In patients with a pre-existing pharyngeal flap, the scope can be passed through one or both ports to visualize the lower vocal tract as long as the ports are not stenosed. A standard speech sample is used at each observation point in the vocal tract. Care must be taken when positioning the scope in the velopharynx. A false positive or false negative diagnosis of velopharyngeal closure will be obtained if the distal end of the scope is not positioned directly above the velopharyn-

geal port. The quality of the study and interpretation, treatment planning, and outcome analysis are enhanced by audio-video recording which allows playback of the study for repeated analysis, comparisons with previous examinations, and demonstration of the findings to the professionals involved in the treatment and the patient and/or parents.

30.8 The Nasometer

The nasometer is a computer-assisted instrument produced by Kay Elemetrics (Pinebrook, New Jersey) which is designed to measure the relative amount of nasal acoustic energy compared to oral acoustic energy during continuous speech production [16]. This instrument uses a sound separator that rests on the patient's upper lip. Microphones on either side of the sound separator sense oral and nasal acoustic energy during speech, and this energy is filtered and digitized by custom electronic modules. The computer with Kay Elemetrics software (Version 1.7) processes the information and produces a "Nasalance Score." This score is a ratio of nasal to oral acoustic energy multiplied by 100. The nasal and oral acoustic energy is averaged during the production of vowels and consonants in test sentences to produce the nasalance score. The length of the speech sample used to calculate the nasalance score may be up to 100 seconds. This instrument is based on the Tonar II developed by Fletcher in 1976 [17]. An abnormally high nasalance score during production of nonnasal consonants suggests velopharyngeal inadequacy and hypernasality, and an abnormally low nasalance score during production of nasal consonants is suggestive of hyponasality and/or nasal airway impairment [16]. Dalston et al. [16] studied the sensitivity and specificity of the Nasometer and found it to be an appropriate instrument for use in corroborating listener judgments of hypernasality. Sensitivity and specificity values for assessing hyponasality were in the expected range for patients without any indication of concurrent velopharyngeal inadequacy, but the scores did not identify hyponasality in patients who exhibited both hyponasality and excessive nasal air emission. Therefore, the instrument may be less useful as a diagnostic procedure in patients who exhibit both velopharyngeal inadequacy and nasal airway impairment [16]. The Nasometer is not a substitute for listener judgments of hyper- and hyponasality but can be useful in providing baseline data to assist in the identification of velopharyngeal inadequacy, assessment of treatment outcome, fitting of a palatal prosthesis, and providing visual biofeedback in speech therapy.

30.8.1 Technique

Care should be taken to ensure that the system is calibrated according to the specifications of the manufacturer. The headgear is then adjusted to fit the patient, and the patient is asked to read or repeat a standard speech sample. This procedure is usually used with patients age 3 and older because younger patients may have shorter attention spans and less well-developed speech and language skills. Once the speech sample is recorded on the computer terminal, the software cursors are used to mark the beginning and end of the speech display. The “calculate” function is then activated, and the mean and standard deviation of the nasalance score are determined.

30.9 Aeromechanical Measurement

30.9.1 Warren and Dubois Technique [18]

Measurements of nasal airflow and of the difference in air pressure above and below the velopharyngeal port may be used to estimate both the area of the velopharyngeal orifice, if any, during the production of stop consonants and the resistance of the port to airflow. Pressure-flow measurements provide information about the coupling of the oral and the nasal cavities during speech and about resistance in the system. They do not describe the movement of particular structures, such as the velum and lateral pharyngeal walls, or the location and configuration of any opening that is present.

30.9.1.1 PERCI

Warren [19] introduced an instrument called the PERCI (Palatal Efficiency Rating Computed Instantaneously) for use in the evaluation of the velopharyngeal mechanism during speech. PERCI records and displays the difference in air pressures in the mouth and in the nose. From a study of 75 cleft palate patients, Warren reported that patients with differential pressure readings >3.0 on the PERCI had velopharyngeal orifice areas of 10 mm^2 or less, whereas those with PERCI readings of <1.0 had areas greater than 20 mm^2 . PERCI readings of 1.0 through 2.9 were associated with velopharyngeal areas between 10 and 20 mm^2 .

30.9.1.2 TONAR

Fletcher and Bishop [17] advanced the study of oral and nasal sound intensity measures as indices to hypernasality through the development of an instrument which they named TONAR (The Oral-Nasal Acoustic Ratio). The instrument prints out voltages associated with the nasal and oral signals and also a trace reflecting the ratio of the voltages from the sound detected in the oral and nasal chambers.

30.10 Summary

Several instrumental procedures are available for assessing the velopharyngeal mechanism and its function. Each has advantages and disadvantages, and choosing among them depends on the specific purpose of the evaluation. The reliability of endoscopic procedures is not well documented.

Aerodynamic measures provide data about the area of the velopharyngeal opening, velopharyngeal resistance to air flow, and air pressure available for the production of obstructed sounds. These measures provide no information about the relative contributions of the velum and the pharyngeal walls to velopharyngeal function.

An important warning in the use of any instrumentation for the study of speech is that data taken during speech production must be interpreted within the context of the patient's repertoire of speech proficiency.

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31 Variations in Nasopharyngeal Skeletal Architecture

SAMUEL BERKOWITZ

31.1 Muscles

31.1.1 Pharynx and Velum [1]

The adult pharynx is the common pathway for food and air in human beings. It extends from the cranial base behind the nasal cavities to the upper end of the esophagus behind the larynx. The pharynx is widest at its upper portion and narrows as it descends to the

esophagus. The pharynx lies immediately in front of the vertebral column, separated from it by prevertebral muscles and fascia. Anteriorly, the pharynx communicates with the nasal cavities, the oral cavity, and the aditus of the larynx. Inferiorly, the pharynx is continuous with the esophagus. The part of the pharynx that extends upward to the level of the velum (soft palate) is commonly called the nasopharynx (Fig. 31.1).

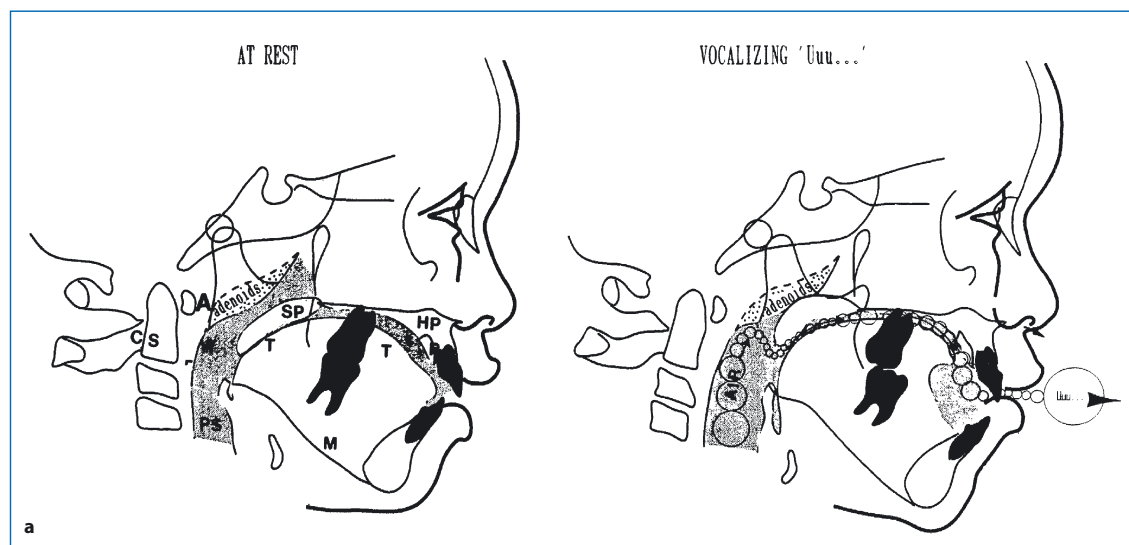


Fig. 31.1. a Lateral cephaloradiograph shows the skeletal structures surrounding the pharyngeal space and allows evaluation of the pharyngeal depth, shape of cervical spine, soft palate size and length, and extent of soft palate elevation. Because it is a two-dimensional representation of the velopharyngeal area and does not show lateral pharyngeal wall motion, this record cannot be used to diagnostically determine velopharyngeal functions. **a Left:** Structures involved in controlling airflow as seen in a lateral cephalograph at 5 years of age (A, anterior tubercle of the atlas; CS, cervical spine; S, odontoid process;

W, posterior pharyngeal wall; PS, pharyngeal space; SP, soft palate (velum); HP, hard palate; T, tongue; M, mandible). **Right:** Air flow when vocalizing “Uuu” during normal speech. The soft palate elevates and makes contact with the adenoids (if present) or the posterior pharyngeal wall during normal speech and swallowing. When the lateral pharyngeal muscles operate in coordination with a competent (adequate length, width, and timing of action) soft palate in a normal skeletal environment most of the air is channeled through the mouth while some enters the nose

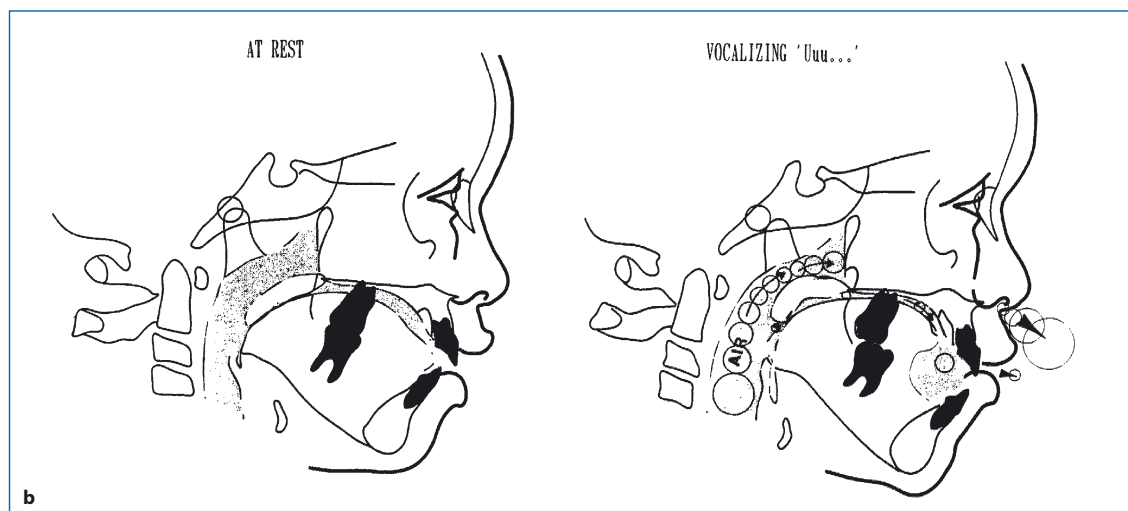


Fig. 31.1. (continued) This is designated as velopharyngeal competency (VPC). **b** Velopharyngeal Incompetency (VPI). There are usually many reasons for inadequate air flow control. Some are: (1) a relatively deep pharynx when related to velar

length; (2) inadequate velar elevation and/or pharyngeal wall motion (neuromuscular function); and (3) poor timing of speech with pharyngeal and velar muscle (sensory-motor) function

The pharyngeal orifices of the auditory tubes open into the lateral walls of the nasopharynx. The auditory tube connects the pharynx with the middle ear and serves to maintain an equilibrium of air pressure between the middle ear and the external atmosphere. The superoposterior part of the pharynx contains the pharyngeal tonsil (also called the adenoid).

The velum (soft palate) forms the boundary separating the nasopharynx from the oropharynx. Because the velum is mobile, this boundary is arbitrary. The velum is a muscular body that attaches to the posterior rim of the hard palate and the lateral walls of the posterior part of the oral cavity. The posterior free border of the velum hangs into the oropharynx and ends in a small midline projection, the uvula.

The oropharynx extends inferiorly to the level of the hyoid bone. The anterior wall of the oropharynx, inferior to the opening into the oral cavity, is formed by the posterior surface of the root of the tongue.

The palatopharyngeal fold (posterior faucial arch) extends from the sides of the velum into the lateral walls of the pharynx. This muscular fold forms the greatest lateral constriction between the oropharynx and oral cavity. The superior end of the epiglottis extends superiorly from the larynx into the oropharynx immediately posterior to the root of the tongue.

The nasopharynx and auditory tube are lined by ciliated, pseudostratified columnar epithelium rich in goblet cells and glands that secrete mucus. The pharynx is lined with stratified squamous epithelium

where contact between the pharynx and velum takes place and in the portion that serves as a food channel.

31.2 Nasopharyngeal Growth

Variations in the height and depth of the nasopharynx are apparent from birth through the early years of growth and development. Rosenberger [2] made a longitudinal study of naso-respiratory areas in children from 3 months to 5 years of age. It was his opinion that an enlargement of the naso-respiratory area accompanied growth in the body and great wing of the sphenoid as well as the forward drift of the hard palate. Brodie [3] demonstrated that the hard palate moves downward, away from the base of the skull, in a parallel manner with increasing age and that this results in a progressive increase in the height of the nasal and nasopharyngeal areas.

In a study of the growth of the pharynx, King [4] concluded that there is a considerable increase in the vertical dimension of the pharynx resulting from the descent of the hard palate, mandible, and hyoid bone, as well as from an increase in the height of the cervical vertebrae. He concluded that there is no appreciable increase in the anteroposterior dimension of the pharynx after an early age.

According to Scott [5–7] growth in the height of the pharyngeal area is regulated by the cartilages of the cervical vertebrae. He, as well as Todd and Tracy [8],

attributed growth in the anteroposterior dimension of the nasopharynx to growth at the spheno-occipital synchondrosis. Subtelny [9, 10] studied vertical growth of the nasopharynx and observed a steady rate of increase up to 15 years of age. Using a measurement from the posterior nasal spine to the soft tissue of the posterior wall of the pharynx, he observed a general increase in depth of the nasopharynx at the level of the palatal plane. Berkowitz [11] believes that this increase is due mainly to a change in the curvature of the posterior pharyngeal wall.

Several investigators studying cleft palate subjects have focused attention on skeletal structures closely related to the nasopharynx. Ricketts [12] found that the basilar portion of the occipital bone varies in its position in relation to the anterior cranial base, and that these variations can influence the anteroposterior dimension of the nasopharynx. Brader [13] reported significantly smaller anteroposterior and vertical nasopharyngeal dimensions in cleft palate subjects than in noncleft individuals and noted that his cleft palate subjects had relatively larger masses of adenoid tissue within the nasopharynx than noncleft individuals. He also observed no significant difference in the angularity of the cranial case in individuals with cleft palate when compared with individuals without clefts.

Moss [14] studied malformations of the skull base associated with cleft palate deformities and indicated that a flexion of the cranial base (N-S-Ba) placed the basioccipital bone “more anteriorly relative to the sphenoid bone,” thus creating a smaller anteroposterior nasopharyngeal dimension. Subtelny [9, 10] showed that width of the skeletal nasopharynx was larger in persons with cleft palate than in normal individuals.

Coccora et al. [15] studied nasopharyngeal growth in children with cleft lip and cleft lip and palate using serial cephalometric films on 57 subjects from 3 months to 7 years of age. Comparisons were made with a noncleft group and with narrative data supplied by Subtelny. Results showed that: (1) The cleft palate group had smaller nasopharyngeal height measurements than the normal group, but only up to 3 years of age. (2) By 3 years of age, all three groups had attained approximately 80% of their respective increases in nasopharyngeal height. (3) Nasopharyngeal depth in the cleft palate group increased slightly more than in the noncleft group (6.8 mm vs. 4.3 mm). (4) The horizontal nasopharyngeal changes in the cleft palate group were shorter than in the noncleft group for most of the age levels studied.

31.3 Functions

The pharynx serves as an air passage connecting the mouth and nose to the lungs via the larynx and trachea and as a food passage connecting the mouth to the stomach via the esophagus. Because these channels are common over part of their length in humans, a sphincter is found at the level of the velum to prevent food from entering the nasal cavities during swallowing.

31.3.1 Swallowing

Constriction of the pharyngeal walls results in changes in the diameter of the pharynx. These changes are important to both swallowing and speech. During swallowing, the lateral and posterior walls of the pharynx move anteromedially, reducing the cross-sectional area of the pharynx. This results in a wave-like constriction that flows down the pharynx and continues throughout the entire digestive tract. The wave-like motion is called peristalsis.

In the initial stage of swallowing, the pharynx constricts around the velum while the velum moves somewhat dorsally and superiorly, closing off the nasopharynx from the oropharynx (a movement called velopharyngeal closure). The oropharynx expands somewhat, and the larynx moves superiorly and ventrally, opening the pharynx to receive the bolus of food. As the bolus moves into the pharynx, the pharyngeal walls constrict behind it, propelling the bolus toward and into the esophagus.

31.3.2 Speech

Oropharyngeal constriction and changes in length also occur during speech. These changes affect the resonant frequencies of the vocal tract, and thus change the quality of the voice and influence the perception of what phoneme is produced. The lateral walls of the oropharynx move medially on low vowels and laterally on high vowels.

Velopharyngeal closure for speech eliminates the nasal cavities from the resonance system and permits the build-up of intraoral breath pressure to produce fricative and plosive consonants. The mechanics of velopharyngeal closure for speech are quite different from those for swallowing. During speech, the velum moves superiorly and posteriorly against the posterior pharyngeal wall (Fig. 31.2), while the lateral pha-

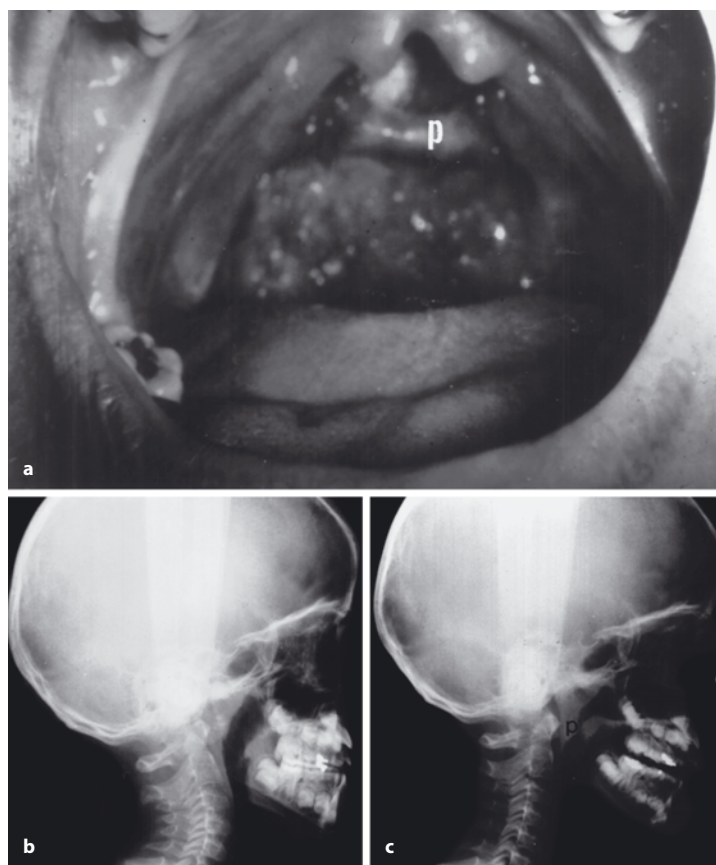


Fig. 31.2 a–c. Passavant's Pad. The superior border of the superior constrictor pharyngeus muscle on contraction forms a ridge or pad on the posterior wall. It is observed in both noncleft and cleft patients. There is no consistent relationship of the pad to the velum and it is usually seen during swallowing as well as during speech, whistling, and blowing. Some state that it functions adequately for compensatory purposes and does not appear to contribute to velopharyngeal closure in all patients. **a** An intraoral view of Passavant's pads (*p*). **b** Lateral cephaloradiograph – at rest. **c** While phonating “Uuu,” showing Passavant's pad (*p*) at the level of the anterior tubercle of the atlas making contact with the uvulae

ryngeal walls move medially against the sides of the velum (Fig. 31.3). By contrast, velopharyngeal closure for swallowing is primarily pharyngeal.

Radiological studies have demonstrated that, during closure for speech, the velum also increases in length, particularly in its posterior part. The velum contacts the posterior wall just above the level of the anterior tubercle of the atlas and on a plane with the hard palate. Velar elevation is somewhat greater for males than for females; and the greatest velar motion is at the junction of its middle and posterior thirds.

It is of interest to note that both velar elevation, and the degree of motion of the lateral nasopharyngeal walls during speech, are greater for high vowels than for low vowels. This is the reverse of the activity of the lateral walls of the oropharynx. The degree of motion, and in fact the degree to which complete closure is achieved, depends on both the phoneme being produced and its phonemic context. Motion is greatest for fricatives, less for high vowels, and least for low vowels. The velopharyngeal function is greater for vowels in a nonnasal context than in a nasal consonant con-

text. Finally, the extent of lateral wall motion and the degree of velar motion are synchronized and directly and highly related to each other.

Velopharyngeal closure during speech has received considerable attention, and properly so because it is an important factor in the treatment of cleft patients. It is evident that the tensor veli palatini muscle is not active in velopharyngeal function. The palatopharyngeus is active during velar lowering, but its activity is inconsistent and vowel-dependent. Palatoglossus activity relates more to tongue motion than to velar activity. The superior constrictor and levator veli palatini muscles are the only two velar muscles studied (the uvulus muscle has not been definitively studied) that are active during velopharyngeal function for speech. All investigators agree that levator veli palatini activity is synchronized with velar height.

Indirect evidence indicates that the uvulus muscle plays an essential role in velopharyngeal function. Nasopharyngoscopy has revealed a patient population with hypernasality, a midline longitudinal concavity rather than convexity of the dorsal velar surface, translucency of the longitudinal velar midline, and a

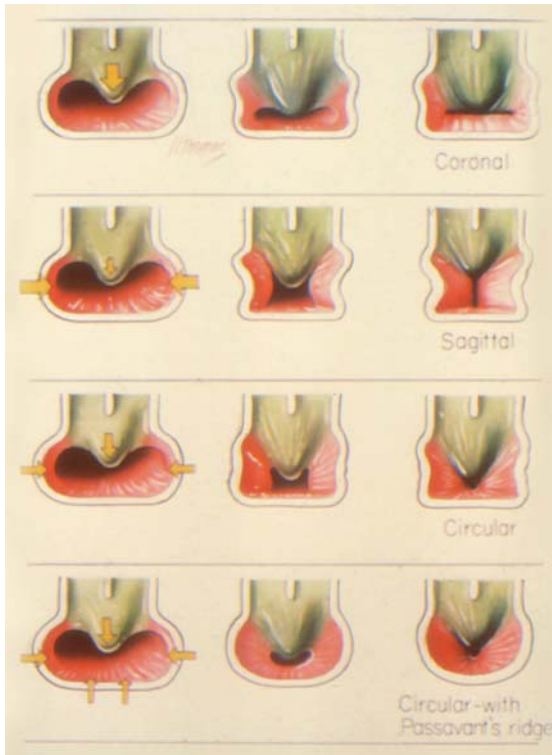


Fig. 31.3. Four basic velopharyngeal closure patterns. The size and shape of velopharyngeal valving patterns is very variable [54]. Although the closure patterns are not actually discrete, for convenience sake, Skolnick et al. [55] categorized velopharyngeal valving into four patterns. These patterns are: Coronal pattern: The majority of valving is palatal with the full width of the velum contacting the posterior pharyngeal wall. The lateral walls move medially to the lateral edges of the velum. There is no motion in the posterior wall. Sagittal pattern: The majority of valving is pharyngeal. The lateral walls move to the midline and approximate each other. The soft palate does not contact the posterior pharyngeal wall, but instead abuts up against the approximated lateral pharyngeal walls, thus completing the closure pattern. Circular pattern: There is essentially equal contribution from the velum and lateral pharyngeal walls with the bulk of the musculus uvulae acting as the focal point. The dorsum of the musculus uvulae contacts the posterior wall (which does not move). The lateral walls squeeze around the bulk of the musculus uvulae. Passavant's ridge pattern: As in the circular pattern, there is essentially equal contribution from the velum and lateral pharyngeal walls, but in addition, the posterior pharynx moves forward. The musculus uvulae also serves as the focal point for closure in this pattern. (Reproduced with permission from [56])

small midsagittal gap between the velum and posterior pharyngeal wall during velopharyngeal closure for speech. This lends presumptive evidence that what has been called the levator eminence is actually a function of the uvulus muscle. In lateral projection, the velum may seem to touch the posterior pharyngeal wall, even when there is actually a small central gap because of the absence of the musculus uvulae.

Thus, velopharyngeal closure for swallowing is accomplished by the superior constrictor, levator veli palatini, and uvulus. Closure for speech is accomplished by the levator veli palatini and uvulus. The difference in function during swallowing and speech is also apparent among persons with velopharyngeal insufficiency due to congenital disproportion between the pharyngeal depth and velar length or disproportion following surgery for cleft palate. In our experience, most individuals in this group who are unable to achieve velopharyngeal closure for speech do so consistently during swallowing.

31.4 The Role of the Nasal Cavity (Fig. 31.4a)

The nasal cavities are the first points of entry and the last points of exit of airflow to and from the lungs. They serve to filter the air as it enters the airway and to control air temperature and humidity. They also serve as resonators for nasal sounds that are produced with the velopharyngeal valve open. The two nasal cavities are separated from each other by the nasal septum in the midsagittal plane. The nasal septum is composed of three parts. The superior part is the perpendicular plate of the ethmoid bone. The inferior and posterior parts are composed of the vomer bone. The anterior part is composed of the cartilage of the nasal septum.

The roof of the nasal cavities is formed by the cribriform plate of the ethmoid bone, which is penetrated by the olfactory nerves. Immediately posterior to the perpendicular plate of the ethmoid is the body of the sphenoid, which contains the sphenoid sinus. The floor of the nasal cavities is formed by the hard palate. Anteriorly, the nasal cavities are bounded by the external nose. The framework of the external nose includes the nasal bones and the nasal cartilages. The bridge of the nose is formed by the nasal bones. Inferiorly, the nasal bones articulate with the lateral nasal cartilages. The tip of the nose is supported by the greater alar cartilages. Small lesser alar cartilages support the lateral walls of the anterior nasal openings.

The lateral wall of each nasal cavity is convoluted owing to the presence of the superior, middle, and inferior nasal conchae. The conchae have an extremely rich blood supply and provide a large surface area for humidity and temperature control of the air passing over them.

Although the terms VP “incompetence,” “inadequacy,” and “insufficiency” historically have been used interchangeably, they do not necessarily mean the same thing. For this reason, standardization of nomenclature has been recommended. Trost-Cardamone [16] has proposed a taxonomy for VP disorders, based on causative factors in which “velopharyngeal inadequacy” is the generic term used to denote any type of ab-

normal velopharyngeal function.” In the broad group of inadequacies, there are subgroups of structural (VPI), neurogenic (VP incompetence), and mislearning (VP mislearning) or functional origins. VPI includes any structural defects of the velum or pharyngeal walls at the level of nasopharynx with insufficient tissue to accomplish closure or some kind of mechanical interference with closure. The all-encompassing term “velopharyngeal dysfunction (VPD)” does not assume or exclude any possible cause of the perceived speech symptoms or management approach. It applies to speech disorders that may be the result of structural deficits, neurologic disorders, faulty learning, or a combination of sources [17].

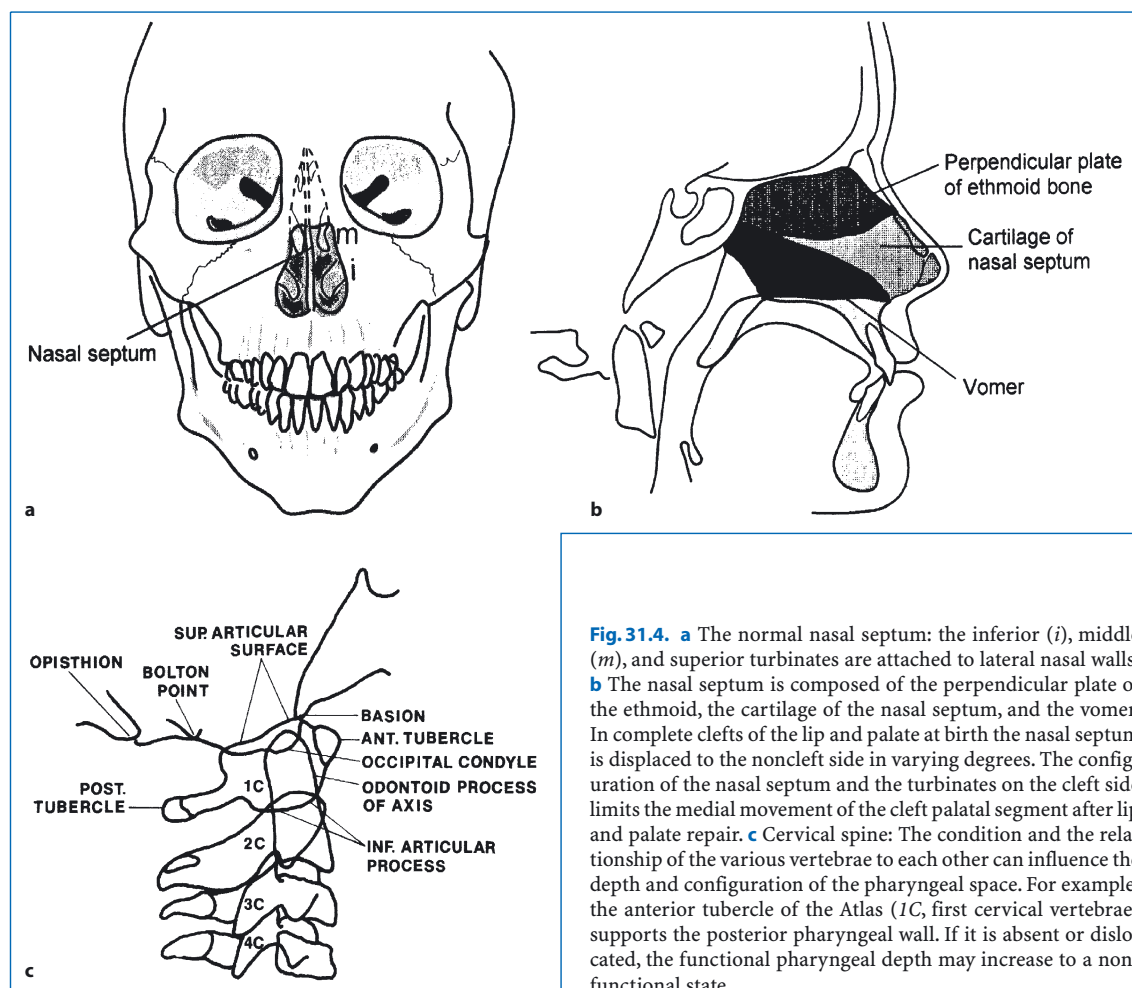


Fig. 31.4. **a** The normal nasal septum: the inferior (*i*), middle (*m*), and superior turbinates are attached to lateral nasal walls. **b** The nasal septum is composed of the perpendicular plate of the ethmoid, the cartilage of the nasal septum, and the vomer. In complete clefts of the lip and palate at birth the nasal septum is displaced to the noncleft side in varying degrees. The configuration of the nasal septum and the turbinates on the cleft side limits the medial movement of the cleft palatal segment after lip and palate repair. **c** Cervical spine: The condition and the relationship of the various vertebrae to each other can influence the depth and configuration of the pharyngeal space. For example, the anterior tubercle of the Atlas (1C, first cervical vertebrae) supports the posterior pharyngeal wall. If it is absent or dislocated, the functional pharyngeal depth may increase to a non-functional state

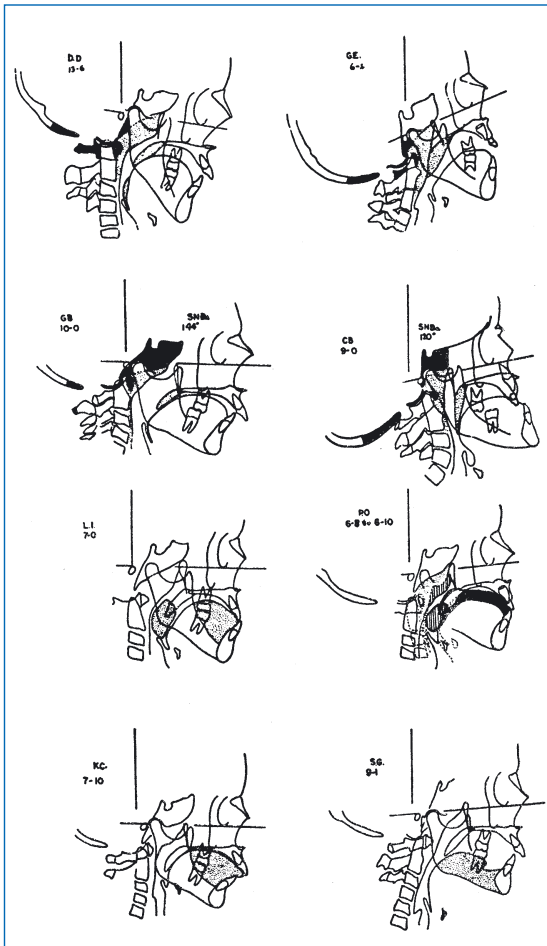


Fig. 31.5. Functional problems related to cervical vertebrae, cranial base, nasopharynx, adenoid, tonsils and the tongue. *DD.*, Dorsal Displacement of vertebrae. *GE.*, Congenital deformity with ventral displacement of cervical vertebrae. *GB.*, Obtuse cranial base upward and backward location of anterior arch of the atlas and severe cleft palate speech. (From [57])

31.5 The Use of Lateral Roentgencephalometrics in Evaluating Skeletal Pharyngeal Architecture and Velar Elevation

Velopharyngeal valving is dependent not only on the sensorimotor adequacy of the velum and its synergistic musculature, but also on the morphologic skeletal dimensions of the nasopharyngeal port. The size and shape of the nasopharynx are determined, in part, by the contiguous osseous anatomy of the maxilla [18], cranial base, and vertebral column (Figs. 31.4, 31.5).

The mechanism of velopharyngeal closure can be studied to some degree by cephalometric roentgenography, but it must be stressed that this is not a functional test for velopharyngeal competency (Fig. 31.6). The vowel /u/, as in “boom,” is generally employed to achieve maximal elevation of the soft palate during vowel production. After careful rehearsal, the subject is instructed to maintain the sound at a constant pitch and intensity for a time sufficient to cover the roentgenographic exposure period. This procedure ensures that the soft palate will remain in a reasonably stable position during the recording interval.

In a noncleft individual, as seen in the lateral head-plate, the soft palate elevates to contact the posterior pharyngeal wall. In doing so, the longitudinal axis of the soft palate becomes continuous with that of the total palatal plane, while the uvula projects downward and nearly at right angles to the rest of the velum. If for any reason this velopharyngeal closure cannot be achieved, deglutition is impaired, and in phonation the air stream is misdirected through the nose. However, it is possible to have good swallowing activity but poor velopharyngeal closure during speech. The term “palatal insufficiency” is more descriptive of a physiologic deficiency than of an anatomic defect.

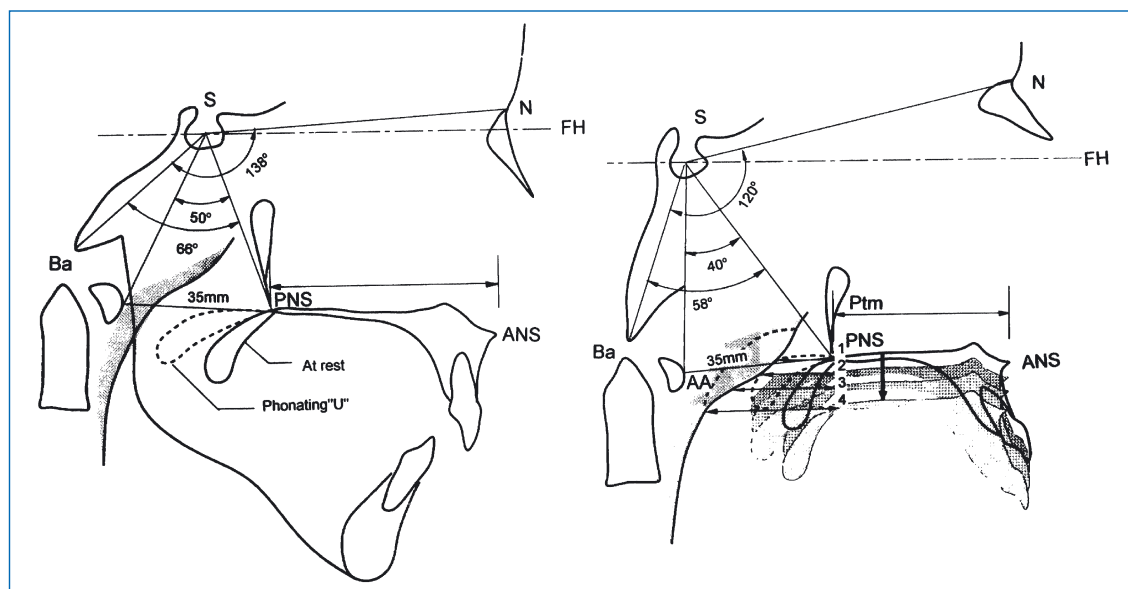


Fig. 31.6. The influence of the skeletal architecture on the pharyngeal form and size. The pharyngeal space is bounded superiorly by the cranial base and laterally by the cervical spine on one side and the tongue/hard palate complex on the opposite side. The odontoid process (axis) of the cervical spine points to the posterior extent (Basion-Ba) of the basilar portion of the occipital bone. It is the median point of the anterior margin of the foramen magnum. The hard palate of the maxillary complex is anatomically associated with the anterior cranial base and can vary in its anteroposterior dimension. The importance of cranial base angle (Ba-S-N) *Left:* Obtuse cranial base angle. In cases with a severe obtuse cranial base angle, the pharyngeal space is usually deeper than normal even in the presence of a long hard palate. An obtuse cranial base positions the cervical spine more posteriorly since it must be associated with the basilar portion of the occipital bone. This condition is usually seen when hypernasality exists in the absence of an overt palatal cleft and is called congenital palatal insufficiency (CPI). *Right:* With an acute cranial base angle, the cervical spine is positioned close to the hard palate creating a shallow pharyngeal

depth. Comment: These relationships change with growth. The palate descends from the anterior cranial base in a parallel fashion. Due to changing slope of the posterior pharyngeal wall, the pharyngeal depth increases with age. The influence of adenoids: The adenoid is attached to the posterior pharyngeal wall above the level of the hard palate. Its size is highly variable according to age. It usually increases until 13 years of age and then retrogresses, but many variations in this growth pattern have been observed. The influence of cleft size: The severity of the cleft size in complete clefts of the lip and palate at birth does influence the nasopharyngeal width before lip and/or palatal surgery but not after the lip is united. The degree of mesodermal deficiency of the hard and soft palate can determine not only the size of the cleft space by influencing palatal size but also the length and width of the soft palate and, therefore, its functional ability to affect velopharyngeal closure (VPC). A short anteroposterior maxillary dimension, as is usually found with submucous clefts, does not by itself signify that VPC will be inadequate. Short anteroposterior hard palate dimension can be associated with a shallow pharyngeal depth

31.6 Cervical Spine Anomalies (Fig. 31.7)

Osborne et al. [19] wrote that fusion of the posterior spines at C2–C3 does not appear to influence the osseous antero-posterior diameter of the nasopharyngeal port. On the other hand, occipitalization of the atlas, a smaller-than-normal anterior arch of the atlas, or atlanto-axial dislocation will have a direct effect on the antero-posterior dimension of the pharynx. It is clear that the population of congenital palatopharyngeal incompetence (CPI) patients with cervical anomalies, of whatever kind, present greater osseous nasopharyngeal depth (the distance between the hard palate and retropharyngeal wall) than do CPI patients without cervical anomalies.

The importance of the anterior tubercle of the atlas and the upper cervical vertebrae in achieving adequate velopharyngeal closure and speech is well-established. One need know only that the musculo-fascial layer covering the upper cervical vertebrae, and forming the posterior pharyngeal wall, is only 2–5 mm thick to realize the morphologic importance of these vertebrae to velopharyngeal closure.

The influence of anomalies of the upper cervical vertebrae on the size and shape of the nasopharynx, with resulting effects on velopharyngeal valving, was first reported by McCarthy [20], and shortly thereafter reemphasized by Schuller [21]. Epidemiologic studies have shown that patients with craniofacial birth defects, many of which are accompanied by velopharyn-

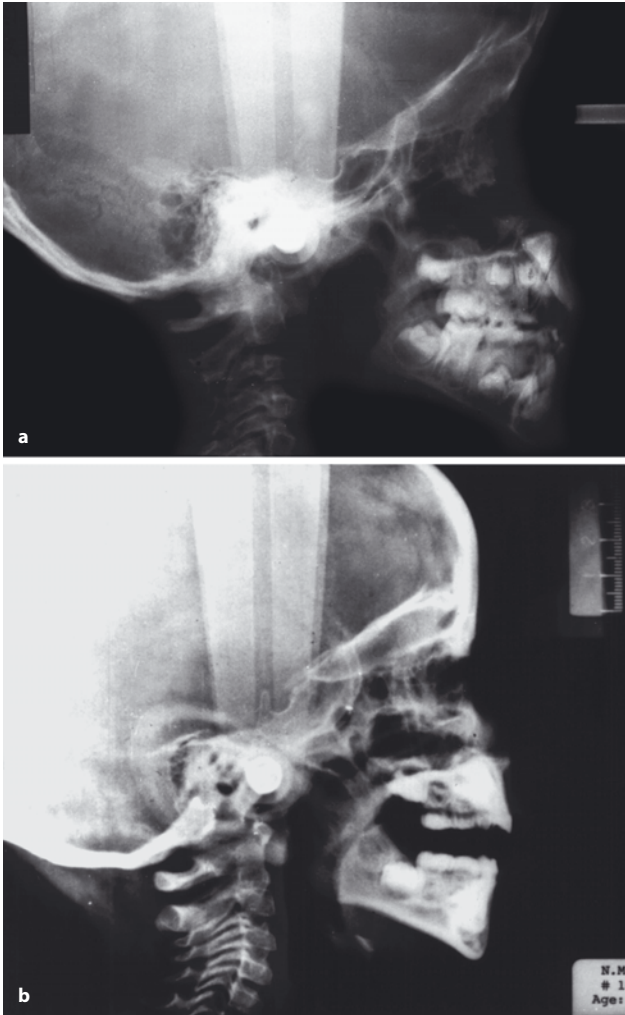


Fig. 31.7 a–e. Cervical spine anomalies and their effects on the pharyngeal space configuration. **a** The absence and **b** malformation or dislocation of the anterior tubercle of the atlas result in the lack of support to the posterior pharyngeal wall, thus increasing the depth of the pharyngeal space. These anomalies coupled with a small velum will compound the problem. *Left: Velum at rest. Right: Velum in function. No contact with the posterior pharyngeal wall*

geal incompetence, have a higher prevalence of upper cervical spine anomalies than the general population [22]. This survey also found that a surprisingly large number (18.8%) of patients with CPI demonstrated anomalies of the upper cervical vertebrae, as discerned in lateral cephalometric head film.

Simply stated, congenital palatopharyngeal incompetence (CPI) can be defined as the presence of hypernasality (rhinolalia aperta) in the absence of an overt cleft palate. Based on detailed studies of 110 subjects, Pruzansky and Mason [23] found that CPI may be

caused by one or more of the following variables: a bifid uvula; a short or thin soft palate, with or without a pink translucent area (“zona pellucida”) usually seen in submucous cleft palate; anomalies of the upper cervical vertebrae and cranial base; and scanty adenoid development or early involution or excision of the adenoid.

As we shall point out in more detail later, our own concept of the defect in congenital palatal insufficiency has been extended to include a broader view of the pharynx and its contiguous structures.

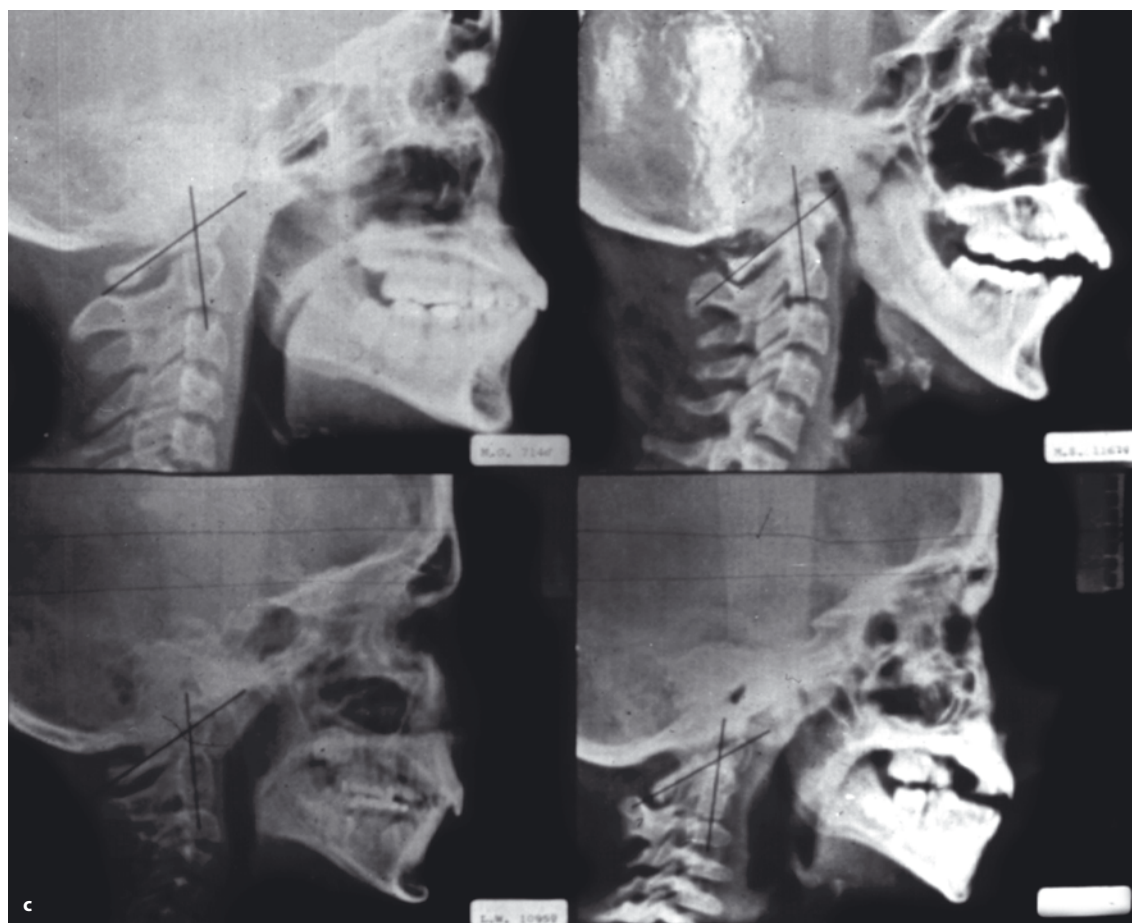


Fig. 31.7 a–e. (continued) Other cervical spine anomalies include: **c** Cervical spine anomalies abnormal atlanto-dental angulation

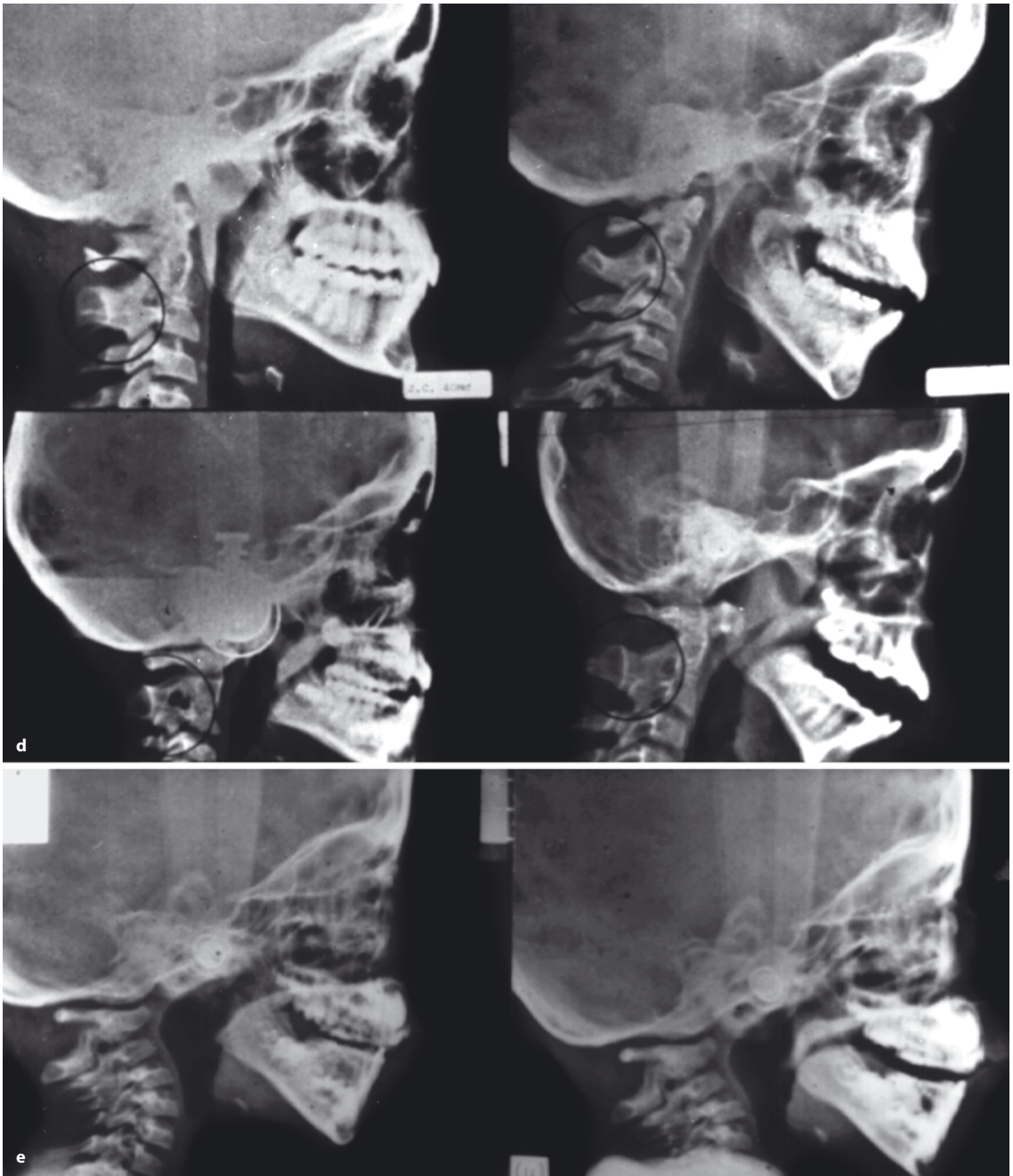


Fig. 31.7 a–e. (continued) **d** Cervical spine anomalies. Fusion of posterior arches C2–C3. **e** Abnormalities to the axis and atlas. (Courtesy of S. Pruzansky)

31.7 Velar Closure

During the clefting process, the muscular forces that act in the region of the pterygoid plates account for the divergence of the pterygoid plates and the consequent increased lateral width of the nasopharynx, the separation of the tuberosities of the maxillae, and the width of the cleft itself. The failure of the hard and soft palate to unite in the midline produces a profound disturbance in the interplay and balance among the involved muscle groups. The tongue, pterygoid muscles, levators, and tensors of the velum tend to aggravate the malformation that has resulted from the failure of union in the midline of the palate.

A more favorable alignment of the osseous segments of the palate will follow re-establishment of balanced muscle forces. Slaughter and Pruzansky [24] concluded that, since repairing the lip altered the spatial configuration of the skeletal framework so profoundly in the anterior segment of cleft, repairing the velum might achieve comparably beneficial results in the region of the nasopharynx.

In reviewing the morphologic variants that might contribute to inadequate velopharyngeal function, a number of factors must be considered. For example, the velum might be too short, the hard palate could be deficient in its anteroposterior dimension, or there may be unilateral or bilateral and posterior pharyn-

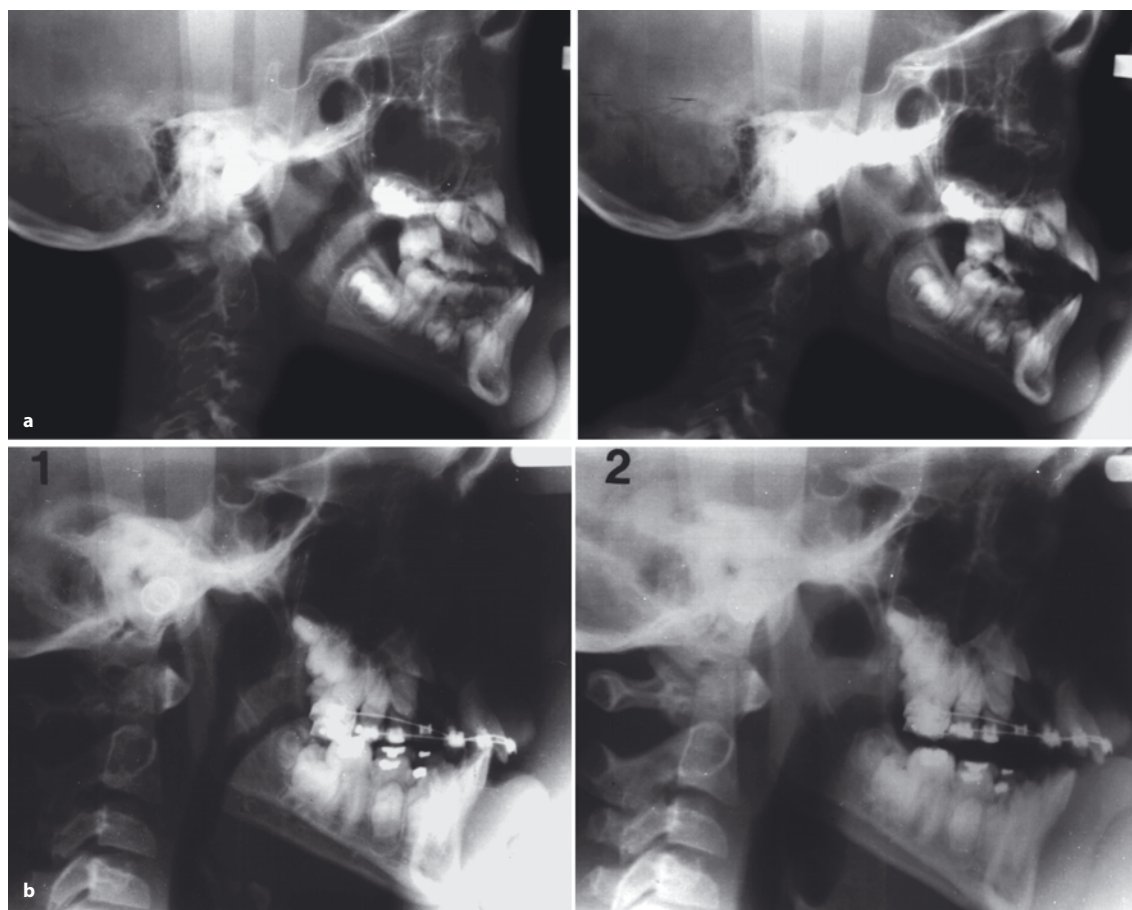


Fig. 31.8 a–e. Variations in the pharyngeal space, velar elevation, and adenoid size. **a** *Left:* Normal pharyngeal architecture at rest, 10 years of age. *Right:* While vocalizing “Uuuu...” Note the velum of good length making contact against a small adenoid – the hard palate is positioned above the anterior tubercle.

b Short, stubby velum in a relatively small pharyngeal space making good contact with the posterior pharyngeal wall at the level of the anterior tubercle, 12 years of age, no adenoids. *1.* At rest, *2.* Vocalizing “Uuuu...”



Fig. 31.8 a–e. (continued) **c** Cephaloradiographs taken at 4 years of age while vocalizing “Uuuu...” The small velum is seen making contact with large adenoid creating a shallow pharyngeal depth. **d** A small, thin paralyzed velum in a deep pharynx at 5 years of age. **e** A long velum of good thickness showing poor elevation resulting in a lack of contact with posterior pharyngeal wall. The velum does not elevate on vocalizing “Uuuu...” The velum may or may not make contact in sustained speech. That is why this record is not a functional test

geal wall dysfunction. In other cases, both the hard and soft palates might be shorter than normal (Fig. 31.8 a–e). In rare instances the soft palate may be paralyzed and incapable of movement (Fig. 31.8f).

The cases presented in Figs. 31.9 and 31.10 show variations in adenoid size and pharyngeal architecture which influence velar pharyngeal closure.

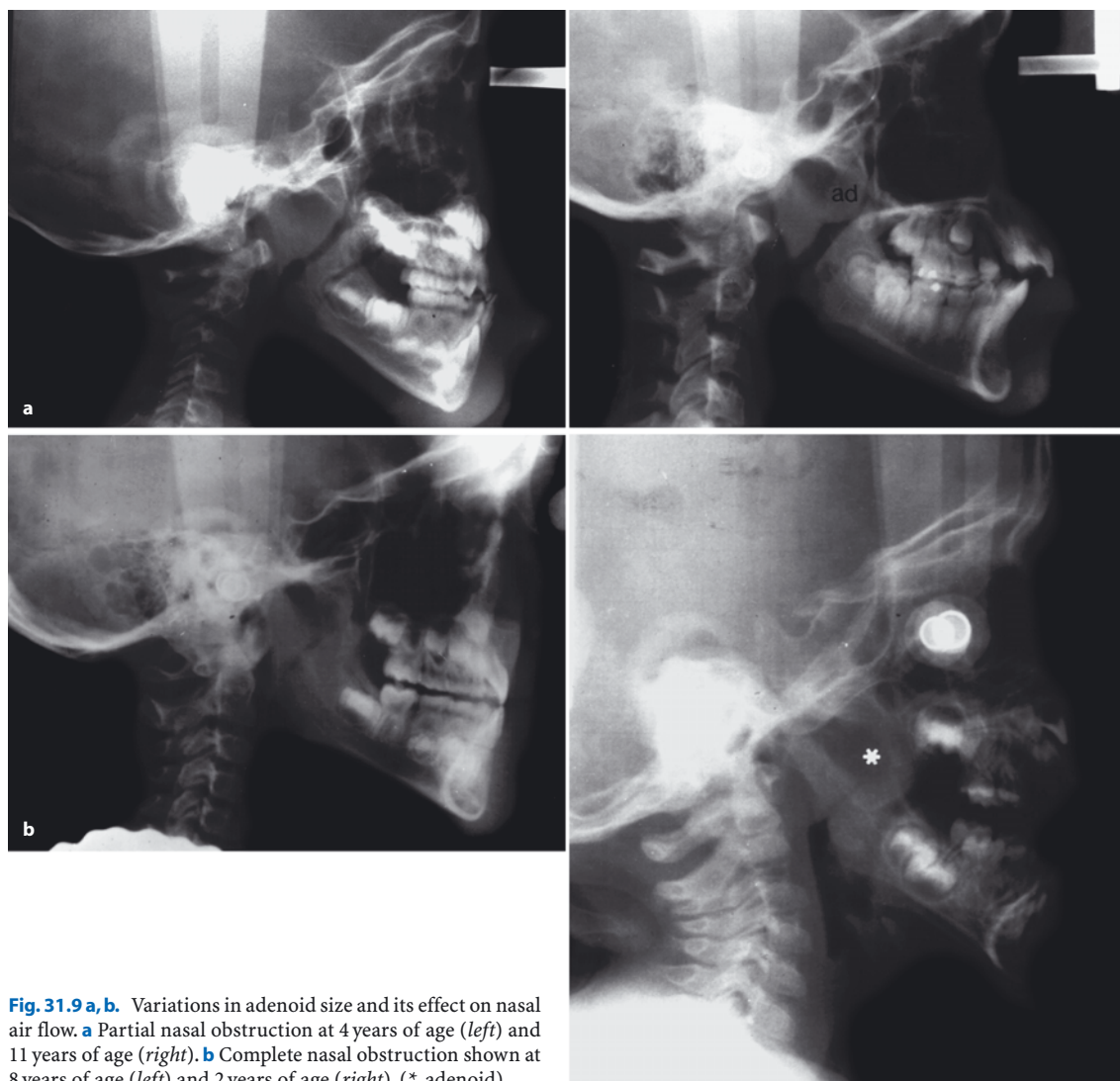


Fig. 31.9 a, b. Variations in adenoid size and its effect on nasal air flow. **a** Partial nasal obstruction at 4 years of age (*left*) and 11 years of age (*right*). **b** Complete nasal obstruction shown at 8 years of age (*left*) and 2 years of age (*right*). (*, adenoid)

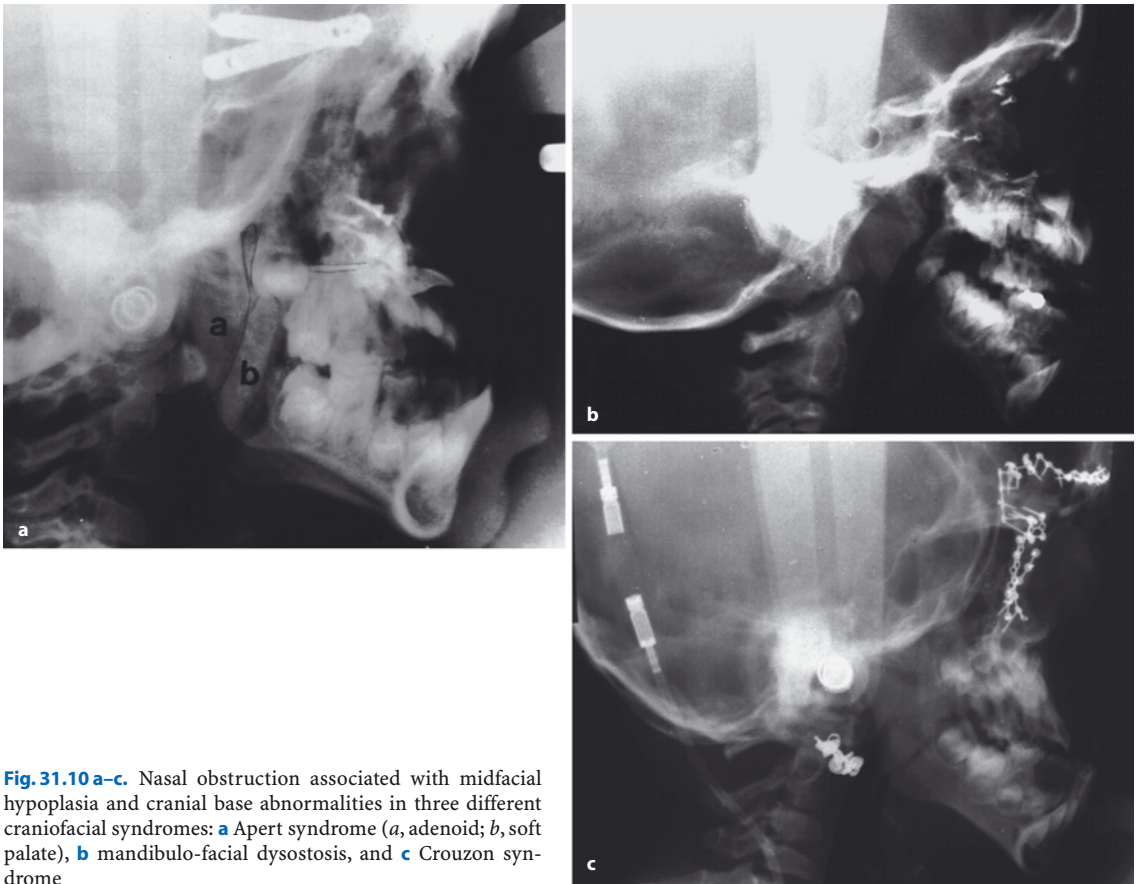


Fig. 31.10 a–c. Nasal obstruction associated with midfacial hypoplasia and cranial base abnormalities in three different craniofacial syndromes: **a** Apert syndrome (*a*, adenoid; *b*, soft palate), **b** mandibulo-facial dysostosis, and **c** Crouzon syndrome

31.8 Improving Velopharyngeal Closure

Traditionally, surgeons have focused on the symptom of hypernasality and have attributed it to inadequate palatal length. Accordingly, operations have been devised to address these structural deficits. Many procedures can be used to control air flow movements. Some have been successful; others have had limited success. The relationships among perceptual, anatomic, and physiologic variables have not been appreciated until relatively recently. To believe that hypernasality generally is the result of inadequate anatomy alone represents a simplistic view of VP function [17].

31.8.1 Pharyngeal Flaps

The pharyngeal flap is the surgical procedure most widely used to correct velopharyngeal insufficiency for speech. One end of the flap remains attached to the pharyngeal wall while the other is sutured to the palate to occlude most of the velopharyngeal space. There are a number of variations of this procedure which involve where the flap from the posterior wall is inserted in the velum. It may be positioned midway on the superior surface, sandwiched (velar split) within the velum, or placed at the end and superior surface of the velum. The posterior flap also may be superiorly or inferiorly based (Figs. 31.11, 31.12). Most surgeons and speech-language pathologists seem to favor the superiorly based flap in which the base of the flap is positioned in line with the anterior tubercle of the atlas. This is the point where the velum in most adults makes contact with the posterior pharyngeal wall during normal speech.

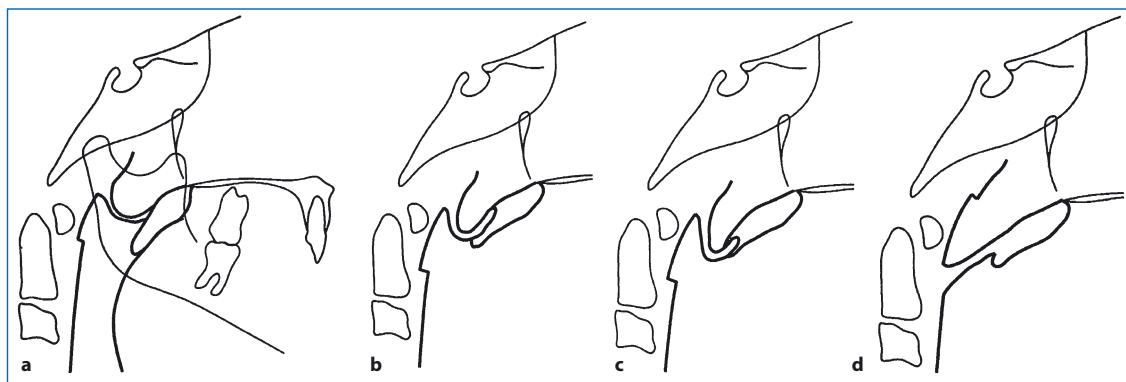


Fig. 31.11 a–d. Variations in the placement of pharyngeal flaps. Superior based flaps are taken from the posterior pharyngeal wall above or at the anterior tubercle of the atlas. **a** Anteriorly superiorly based flap. **b** Posteriorly superiorly based flap. **c**

Velar split – superiorly based. Flap is taken from the pharyngeal wall at the level of anterior tubercle. **d** Inferiorly based flap was taken from the posterior pharyngeal wall below the anterior tubercle of the atlas

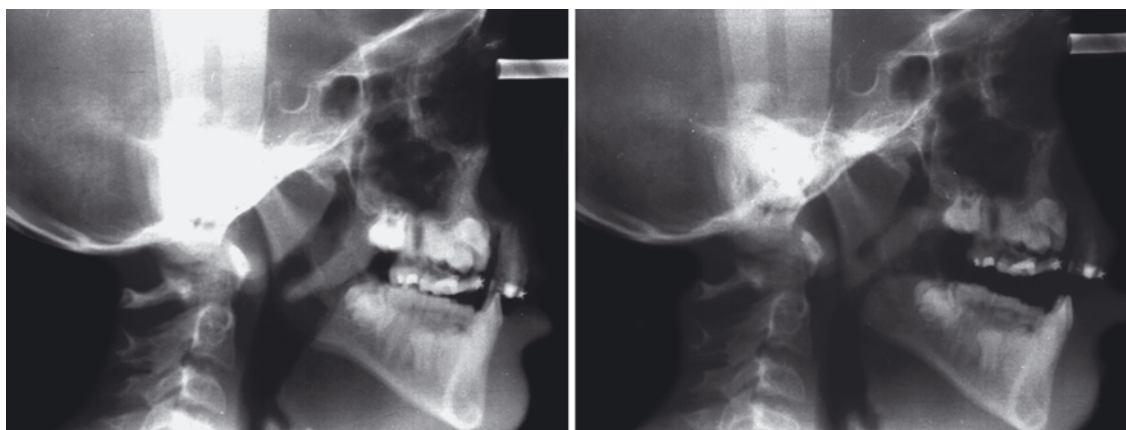


Fig. 31.12. Inferiorly based pharyngeal flap in the presence of a small adenoid. Even with good velar length and elevation, a pharyngeal flap was necessary because of poor lateral pharyngeal wall muscular movement. *Left:* At rest. *Right:* While vocal-

izing “Uuuu...” Inferiorly based flaps and/or flaps placed below the anterior tubercle of the atlas usually do not function adequately because that may not be the position for maximum lateral pharyngeal muscle action

The pharyngeal flap is an unphysiological substitute for the velopharyngeal mechanism because it acts as a sail to capture and channel most of the air through the mouth. The remaining pharyngeal port size will determine whether hypernasality or hyponasality will be present after surgery. Warren [25, 26], analyzing aerodynamic pressure flow patterns in normal speech, determined that hypernasality occurred when the velopharyngeal port exceeded 10 mm^3 and that nasal escape of air was clearly evident at 20 mm^3 . Bjork [27, 28] using basal radiographic techniques, estimated that a port size of 20 mm^3 in area was the threshold between hypernasal and hyponasal speech.

Shprintzen et al. [29] reported that the flap may have to be positioned more to one side than another according to the location of the port space. With recent improvements in diagnostic abilities to determine the location and degree of air escape, nasopharyngoscopy has become the procedure of choice to predetermine the lateral position to place the pharyngeal flap. Unfortunately, surgical skill is not the sole determinant of the procedure's success, and patients still need to undergo extensive speech therapy.

Occasionally, a patient will remain hypernasal and continue to produce articulation errors in spite of a seemingly good surgical result. This can occur even when there seems to be a good speech mechanism, good muscular motion, and a good family environment.

An excessively wide flap can block off too much air flow into the nose, leading to mouth breathing and denasalized nasal consonants, and patients may suffer other side effects ranging from severe snoring to sleep apnea. As in normal speech, the opening and closing functions of the nasopharynx necessary for speech must be achieved by lateral wall movements in the nasopharynx. The lateral walls need to move medially to make contact with the flap during speech to prevent nasal escape of air.

A pharyngeal flap was also used in conjunction with push-back procedures at the time of palatoplasty, thinking that when combined the repositioned velum would be better maintained in its posterior position. Experience has shown that this should never be performed as a primary procedure because there is no evidence that velopharyngeal incompetency can be detected with certainty prior to speech production. Scar contracture resulting from push-back procedures not only inhibits palatal growth but causes any velar length increase to relapse. It does not seem wise to tether the push-back tissue with the pharyngeal flap because negative changes occurring to the palatal tissue can distort the flap.

In the treatment of cleft palate, the superiorly posterior-based pharyngeal flap is almost universally favored. Some clinicians are also using inferiorly based flaps in some cases [30, 31]. It is still debated whether the superiorly or inferiorly based procedure is better. However, Trier [32] points out there are compelling reasons why the superiorly based flap is employed more frequently. The inferiorly based flap not only has severe length limitations but also has the disadvantage of tethering the flap in an inferior direction, away from the palatal plane and motion for affecting VP closure.

Most speech-language pathologists tend to favor performing a pharyngeal flap at 4 years of age to avoid the problems of prolonged training. Early pharyngeal flap surgery carries no systemic risk of interference with facial growth, but can cause some reduction in forward maxillary growth. It also can cause hyponasality and sleep apnea if it significantly obstructs air flow through the nose [33]. The ability of patients to achieve proper velopharyngeal closure after pharyngeal flap surgery is unpredictable. Retrospective studies have shown that, when performed prior to 3 years of age, there are no significant morphological differences in the velopharyngeal area among the various cleft groups. Harding and his colleagues [34] advocated a superiorly based pharyngeal flap at a mean age of 6.5 years. Randall et al. [35] advocated performing the procedure even earlier, between 2 and 3 years of age, especially when associated with a short, scarred, immobile palate with severe hypernasality and virtually no anterior articulation.

Failed pharyngeal flaps are generally due to the inability of the lateral pharyngeal walls to gain firm contact with the sides of the pharyngeal flap. This can result if the flap is too narrow and/or the lateral pharyngeal walls do not move far enough medially to reach it. In some instances, the flap may be set below the point of maximum medial movement, which is usually at the level of the hard palate. He also states that, under no circumstances, should a pharyngeal flap operation be performed simultaneously with other procedures.

31.8.2 Speech Aid Appliances

Wolfaardt et al. [36] noted that the functional integrity of the palatopharyngeal valve can be compromised by a number of factors, including neurologic disorders such as stroke or head trauma; degenerative diseases such as multiple sclerosis, Parkinsonism, or bulbar palsy; craniofacial birth defects, such as overt and covert clefts of the palate; and some behavioral disorders (e.g., functional articulation disorders).

In addition, in the absence of a palatopharyngeal control problem, resonance balance in speech also may be rendered aberrant by hearing loss or faulty sensorimotor learning patterns. These include impairments associated with congenital neurologic disorders (e.g., cerebral palsy) that impair a speaker's ability to regulate the articulatory dimensions of speech that influence the acoustical expression of nasality.

Prosthetic dental appliances are used for the replacement of missing or malformed teeth in the line of the cleft and, when indicated, in the design of speech appliances.

In the event that surgery is contraindicated, delayed, or performed unsuccessfully, a dental speech appliance may provide a satisfactory substitute. Such appliances are usually made of acrylic plastic and retained by clasps about the teeth. Metallic castings of nonprecious and precious metals also may be used in the design of speech appliances.

For purposes of description, the appliance consists of three parts: the (a) palatal, (b) velar, and (c) pharyngeal sections (Fig. 31.13). The palatal section may consist of little more than a covering for the hard palate and carry the clasps for attachment to the teeth. This section may also supply replacements for missing teeth. In cases in which vertical dimension must be increased because of arrest of vertical growth of the middle face, the palatal section may cover the natural teeth. In these circumstances, the teeth should be crowned to protect them from dental caries. By forward extension, the palatal section may also plump the upper lip and improve the facial profile.

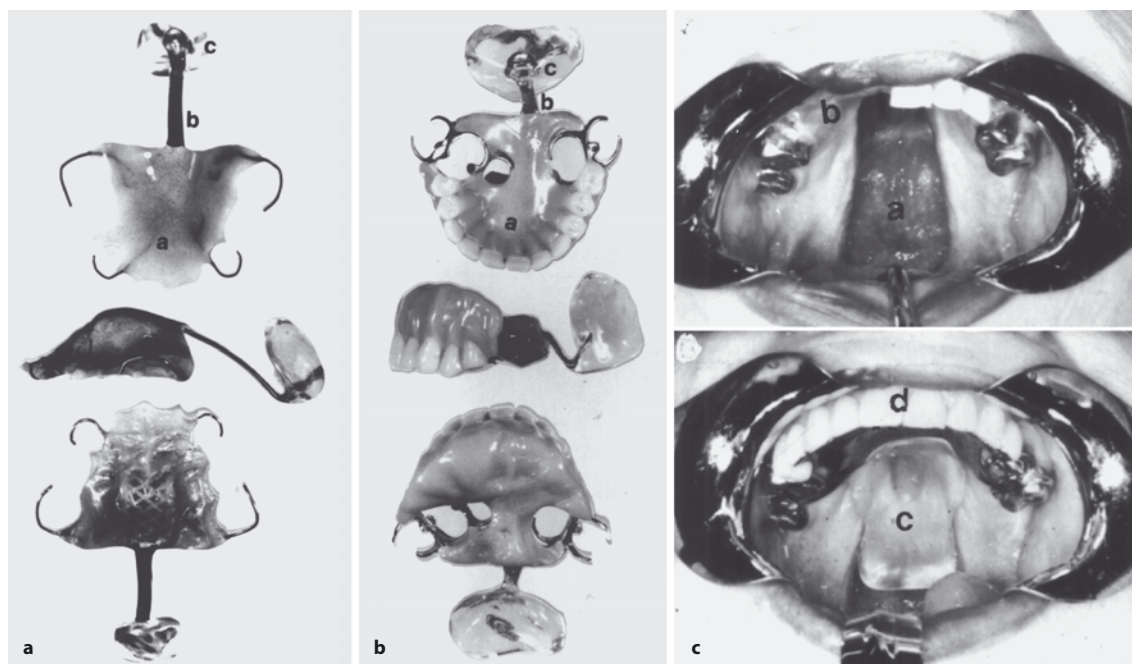


Fig. 31.13 a–c. Speech aid appliances. In the event that surgery is contraindicated or delayed or, if performed, does not produce proper palatopharyngeal closure, a dental speech appliance may be substituted. Such appliances may be made of acrylic plastic retained by clasps about the teeth. Metallic castings of nonprecious and precious metals may also be used. **a** The appliance consists of three parts: (a) the palatal, (b) the velar, and (c) the pharyngeal sections. The palatal section (a) covers the palate and carries the clasps for attachment to the teeth. **b** The palatal section supports placement for the missing teeth and may also plump the upper lip to improve the facial profile. The velar section (b) is in the shape of a bar connecting the palatal

section to (c) the pharyngeal portion of the appliance which extends into the nasopharynx and assists in palatopharyngeal closure and reducing nasal resonance. **c** When midfacial vertical dimension is lost (*top*) and must be increased the palatal section will need to replace missing teeth and cover the natural teeth (*bottom*, wearing appliance). In such cases the teeth need to be crowned in gold to protect them from dental caries. The unoperated cleft space (a) is covered with a posterior extended acrylic section (c) to help control air flow, and permit proper feeding. (*d*) Replacement teeth. The pharyngeal section needs to extend into the nasopharynx and make contact with the posterior pharyngeal wall. (Courtesy S. Pruzansky)

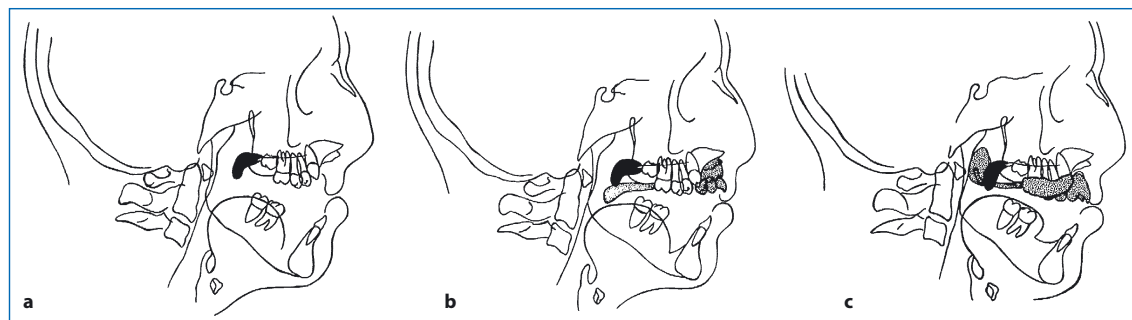


Fig. 31.14 a–c. The pharyngeal bulb in a speech appliance is viewed in cross-section in place in the nasopharynx. **a** Incompetent velopharyngeal closure. **b** Inadequate nasopharyngeal

bulb. **c** Pharyngeal bulb (pharyngeal extension) is well positioned within the nasal chamber. (Courtesy J.D. Subtelny)

Fig. 31.15. The pharyngeal bulb on a speech aid appliance is viewed in the nasopharynx. At rest (*left*), a lateral space on either side of the bulb allows for normal nasal drippings to enter the mouth. When phonating /a/, the lateral pharyngeal walls make contact with the speech bulb, reducing nasal air flow. (Courtesy of J.D. Subtelny)

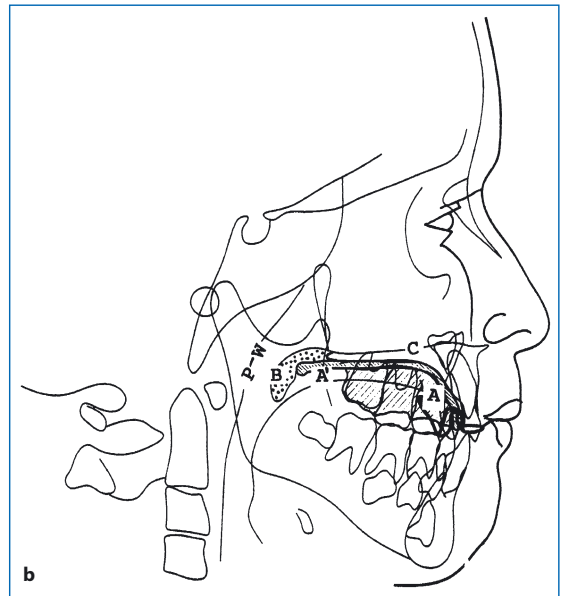
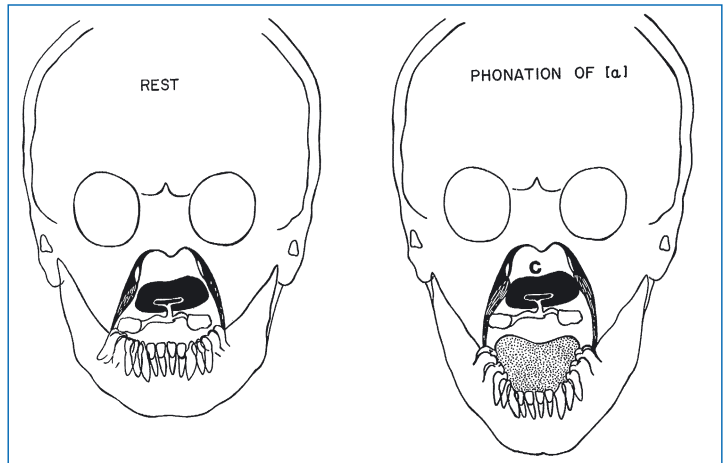


Fig. 31.16 a,b. A palatal lift appliance. It is used to improve velopharyngeal function by improving velar muscle action. **a** The appliance on a dental model. The velar extension extends posteriorly to the uvulae. **b** Lateral cephalometric tracing show-

ing the appliance (A-A') velar section making contact up to the uvulae (B). The appliance can be well stabilized by using orthodontic bands with soldered buccal shelves to establish undercuts for the clasps

The velar section is in the shape of a bar connecting the palatal section to the pharyngeal portion of the appliance. The pharyngeal portion extends into the nasopharynx and assists in palatopharyngeal closure (Figs. 31.14, 31.15). The skill of prosthodontists in designing these devices has added a fortunate dimension to speech habilitation of cleft palate whenever surgery is unable to provide good results.

This treatment technique is logically combined with auditory training. The patient needs to learn to rely on auditory and intraoral somesthetic percep-

tions of resonance balance and palatopharyngeal control. Articulation training is likewise conducted to normalize consonant distortion or substitution patterns that patients may have developed as compensatory maneuvers in the face of palatopharyngeal incompetence.

Some speech-language pathologists and dentists utilize a palatal lift appliance (PLA) which is designed to hold the soft palate up and backward and to ultimately improve its function by acting as an isometric training appliance (Fig. 31.16) [36, 37]. D'Antonio

(personal communication, 1995) suggests that the PLA be used for patients in whom there is adequate tissue but poor control of coordination and timing of VP movements. It is hoped that the degree of lift can be reduced gradually until the appliance can be discarded. Unfortunately, in many instances the prosthesis must be worn indefinitely. The use of a palatal lift appliance (PLA) is still controversial. Some believe that, at best, its use produces inconsistent results even with speech therapy. The palatal lift appliance (PLA) has also proved useful in reducing drooling and improving mastication and tongue movement. Speech therapy associated with use of a PLA may include any one or a number of techniques, many of which are practiced by the patient with and without the appliance in place [36].

Auditory training should be used to enhance the patient's auditory awareness of normal and deviant resonance balance. Articulation training is used to develop a patient's palatopharyngeal control to regulate nasal resonance.

31.8.3 Push-Back Procedures (Velar Lengthening)

These are surgical procedures designed to lengthen the soft palate done either in conjunction with primary palatoplasty or as a secondary procedure after the palatal cleft has been closed and velopharyngeal incompetence has been detected. The most common surgical procedure is the V-Y push-back, commonly called the Wardill-Killner [38, 39] or Veau-Wardill-Killner procedure. There are a number of variations of this basic design. This procedure involves the use of two unipedicle or single-base flaps of palatal mucoperiosteum, the soft tissue on both sides of the cleft space. The flaps are elevated from the bony palate and repositioned so that the cleft space is covered and the length of the soft palate increased. Push-back palatoplasty procedures performed at an early age require an extensive amount of mucoperiosteal undermining and soft tissue displacement leaving large areas of denuded bone. These areas are later covered over by scar tissue which inhibits palatal bone growth and causes palatal deformation. This problem exists even when the push-back procedure is performed as a secondary procedure after the palatal cleft has already been closed.

Lateral cephalometric evaluation of this and other push-back techniques has shown that the net gain in soft palatal length is not as great as the amount of mucoperiosteum brought posteriorly because of contraction of scar tissue. Yet, some speech-language pathologists believe this technique yields a higher success rate, as measured by speech results, than any other

surgical technique. Harding et al. [40] criticized this procedure because large surfaces of denuded bone are left in the anterior palate which result in excessive scarring and midfacial growth retardation. Other palatal lengthening procedures, such as Millard's Island Flap, have not been successful because the extensive mucoperiosteal shifting leaving large areas of denuded bone has been shown to have a deleterious effect on palatal development.

Witt and D'Antonio [17] recommend Furlow's double-opposing Z-plasty for patients with a small central VP gap seen in submucous cleft palate or in patients with previously repaired cleft palate who demonstrate a midline "trough" or muscular diastasis.

31.8.4 Sphincteric Pharyngoplasty (SP) of Orticochea [41, 42]

No discussion of pharyngeal surgery could be complete without mentioning the Orticochea operation. Jackson [43] described the procedure most commonly used today. This surgical procedure was designed to occlude the lateral ports by changing the lower insertion of the posterior pillars from the lateral walls to the posterior wall of the pharynx. Orticochea believed that this created a dynamic muscle sphincter of the pharynx, which could open and close dozens of times a minute. Furthermore, this sphincter has the same cerebrocortical role as the nasopharyngeal sphincter described by Passavant because the palatopharyngeus and superior constrictor muscles that are part of the Passavant nasopharyngeal sphincter are used in its constriction.

In patients with very short palates in whom there is a greater risk of dehiscence, only one of the palatopharyngeus muscles is transplanted. Three months later, in a second operation, the sphincter is completed by transplanting the second palatopharyngeus muscle.

The sphincter is created by transposing the posterior tonsillar pillars with the enclosed palatopharyngeus muscles from the lateral pharyngeal walls to the midsection of the posterior pharyngeal wall. Three openings conduct air between the oral and nasal pharynxes. The central opening's shape and width remain unchanged postoperatively, with no reduction of the lumen. The two lateral openings completely disappear.

This sphincter tends to close off the nasopharynx. Some surgeons use it as a secondary corrective procedure.

Sphincter palatoplasty's cited advantages include: (1) dynamic sphincteric closure as a result of retained neuromuscular innervation, (2) technical ease of execution, (3) a low complication rate, (4) nonobstruction of the nasal airway, and (5) no violation of the

velum. Unfortunately, the question of whether it attains superior speech results is still open. It also must be emphasized that many surgeons have developed variants of the original procedure. The SP procedure has theoretic advantages and is an effective means of management of VPI for some patients. More studies need to be performed to determine when this or the pharyngeal flap should be the treatment of choice [17].

31.8.5 Posterior Pharyngeal Wall Augmentation

Teflon and other materials have been implanted in the posterior pharyngeal wall with no reported long-term success. Potential problems include tissue incompatibility and migration of the implant. Injectable forms of these substances also can create problems with respect to embolism or transport of the prosthetic matter to regional lymph nodes.

Although pharyngeal flap surgery is the treatment of choice in most cleft palate patients, Denny et al. [44] believe that a posterior pharyngeal wall implant of costal cartilage should be used in cases of extremely shallow pharyngeal spaces with inadequate lateral pharyngeal wall motion and when the side effects of a pharyngeal flap should be avoided. Shprintzen et al. [45] and Sher et al. [46] believe that pharyngeal flaps should be avoided in patients with primary neuromuscular disease or pharyngeal hypotonia.

Pharyngeal implants have been carried out with a variety of materials by a number of surgeons over the years (e.g., Teflon, by Smith and McCabe [47]; Silastic by Blocksma [48]; Silicone Gel by Brauer [49]; Proplast by Wolford et al. [50]; and Homologous Cartilage by Trigas et al. [51]).

Surgical procedures as well as injection techniques have been used. Furlow et al. [52] reported one instance of obstructive sleep apnea (OSA) 6 years post-Teflon injection due to the downward displacement of the Teflon particles resulting in a narrowing of the patient's airway. In a series of studies, Vinas and Jager [53] obtained the best results with surgically inserted cartilage implants.

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32 The Velopharyngeal Mechanism

ROBERT J. SHPRINTZEN

Normal speech is dependent on the modulation of voice and sound and the appropriate direction of air-flow from the lungs as it travels through the vocal cords and the upper airway. One of the most important components of speech is the ability to separate the oral cavity from the nasal cavity, a process that is unique to humans [1]. The majority of speech messages are delivered by the production of consonants [1], and the large majority of consonants are produced orally with no nasal resonance of sounds. The only consonant sounds that have nasal resonance in English (and most other languages) are /m/, /n/, and /ng/. All other consonant sounds require the separation of the oral cavity and nasal cavity accomplished by the movements of the velum and pharyngeal walls referred to as velopharyngeal closure. Failure to achieve velopharyngeal closure has been variously referred to as velopharyngeal incompetence, velopharyngeal insufficiency, velopharyngeal inadequacy, and velopharyngeal dysfunction. Arguments over the validity of any or all of these terms are not really important and for the purpose of this chapter, we will simply refer to the problem as VPI, the most widely used and accepted term historically.

32.1 Normal Velopharyngeal Closure

The opening between the oral cavity and nasal cavity is demarcated anteriorly by the velum, posteriorly by the posterior pharyngeal wall, and laterally by the lateral pharyngeal walls, all muscular structures that normally move during swallowing and speech. This opening, sometimes referred to as a “port” or “portal” is really a tube oriented in a relatively vertical manner (Fig. 32.1) [2]. The opening and closing of this pas-

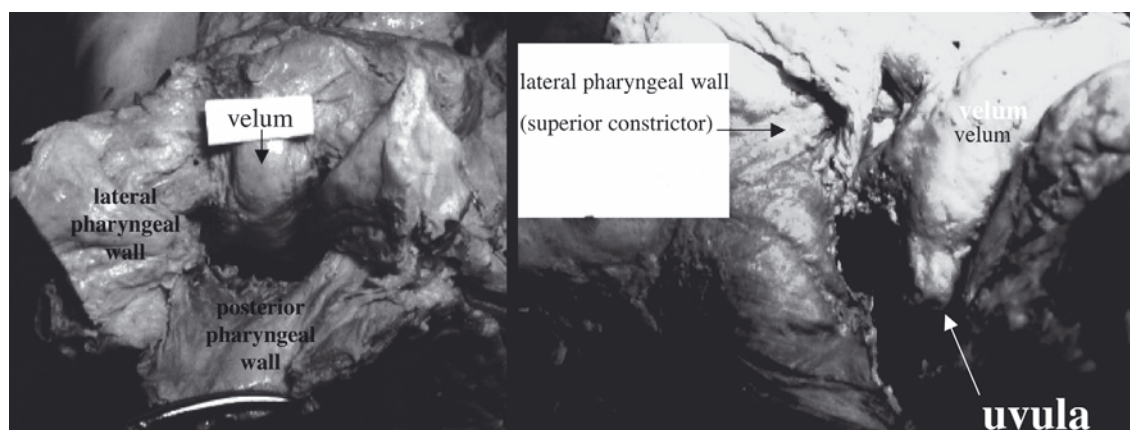


Fig. 32.1. Two views of the anatomy of the velopharyngeal valve. At *left* is an axial view from above showing what looks like a two-dimensional opening, but when the same specimen is viewed from a different angle (from the right posterolateral

aspect of the pharynx), the vertical height of the velopharyngeal mechanism can be appreciated. Therefore, the velopharyngeal valve is a three-dimensional tube with width, breadth, and height

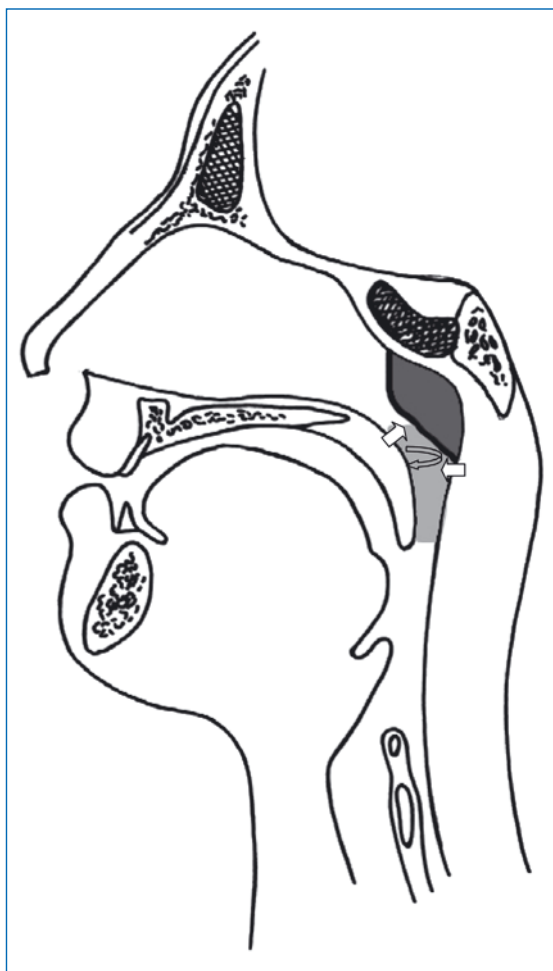


Fig. 32.2. The planes of movement necessary for velopharyngeal closure

sageway between the oropharynx and nasopharynx actually represents a valve or valving mechanism, similar to other valves in the body that regulate the flow of air, blood, or other fluids by modulating the size of the opening. The movements for speech in the velopharyngeal mechanism are controlled by the motor cortex and are learned, just as the movements of the tongue, lips, and teeth are learned for speech. The movements of the same structures during swallowing

are primarily reflexive and mediated by the brainstem. Velopharyngeal movements for swallowing are unrelated to those of speech, and the muscles used and timing of movements are different [3]. For the velopharyngeal valve to close during speech, there needs to be movement from the velum and the pharynx along three planes: the antero-posterior plane, the medial plane, and the vertical plane (Fig. 32.2). Antero-posterior movement is caused by the contraction of the levator veli palatini muscle which acts like a sling to draw the velum back towards the posterior pharyngeal wall. Depending on the individual craniofacial structure of the patient, that movement can be primarily posterior, primarily superior, or postero-superior (Fig. 32.3). The medial movement of the pharyngeal walls is caused by contraction of the uppermost fibers of the superior constrictor pharyngeus. Lateral pharyngeal wall motion is highly variable ranging from barely detectable motion to lateral pharyngeal walls that approximate in the midline of the pharynx. Anterior motion of the posterior pharyngeal wall, usually referred to as Passavant's ridge, is also caused by contraction of the superior constrictor and can be variable in shape, size, and position (Fig. 32.4).

The variability of movement in the velopharyngeal valve from person to person was first described in a landmark article by Skolnick and colleagues in 1973 [4]. They showed that some individuals have more lateral pharyngeal wall motion than others, some have Passavant's ridge and some do not, and that this variability of movement is true for both normals and people with velopharyngeal insufficiency. Subsequent research in large samples of both normals and people with VPI confirmed this finding [5]. This variability may be related in part to structural differences from person to person, but learning is also clearly a factor. The role of learning has been demonstrated experimentally by using visual nasopharyngoscopic biofeedback to alter patterns of movement [6] therefore indicating that it can be altered by learning new muscular movements.

Therefore, velopharyngeal closure is a voluntary, CNS-mediated motor activity that is learned during early speech acquisition. The role of diagnosis needs to be aimed at determining if a failure of the velopharyngeal mechanism to close completely during normally nonnasal speech is an anatomic or physiologic limitation versus an error in learning.

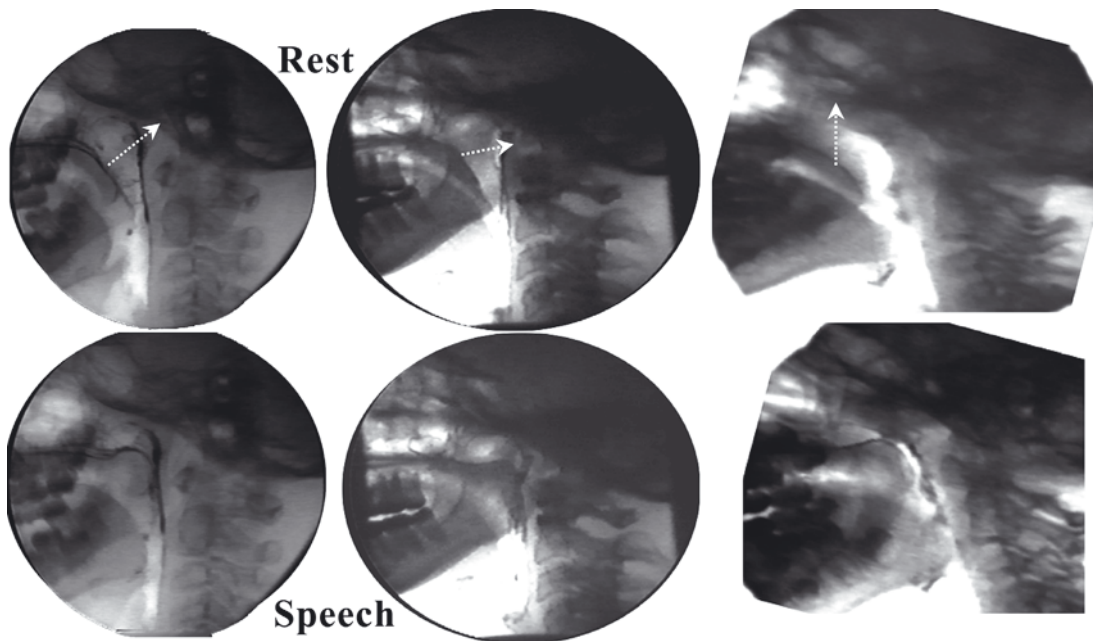


Fig. 32.3. Three lateral view videofluoroscopies showing the different types of movement of the velum that can occur depending on the orientation of the levator palatini

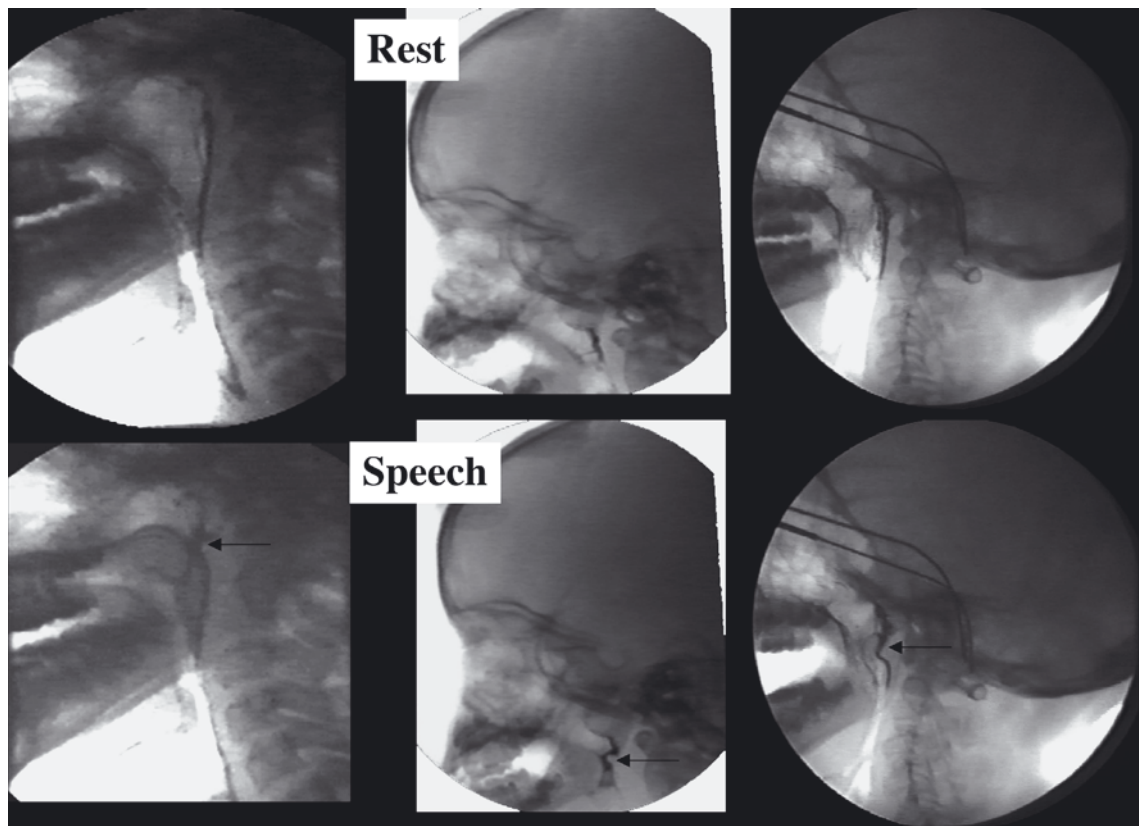


Fig. 32.4. Several different examples of Passavant's ridge (arrow)

32.2 Failures of Velopharyngeal Valving

There are a substantial number of reasons why velopharyngeal closure might not occur during speech that normally requires it. VPI is usually associated with palatal anomalies, but the frequency of VPI in disorders unrelated to structural malformations of the palate are also very common. There are no definitive data available that help us to determine what the most common cause of VPI might be, but suffice it to say that it is a common disorder. For the purposes of this text, we will focus primarily on clefting anomalies, but the reader should be aware that problems with the velopharyngeal valve associated with other disorders requires careful scrutiny as well.

Disorders of velopharyngeal function might best be divided into three major categories: structural, neurogenic/myopathic, and learned. These categories are not mutually exclusive. In other words, it is typical to have only one dysfunction causing the problem, but it might also be possible to have two or three of these factors in operation simultaneously.

32.2.1 Disorders of Structure

Structural anomalies include clefting of the palate (in all of its variations), anomalies of symmetry of the palate and/or pharynx, and anomalies of the surrounding pharynx (including abnormalities of the lymphoid tissue). Cleft palate is a broad designation for a broad spectrum of anomalies that range from complete clefts of the lip and palate to a nearly undetectable malformation known as occult submucous cleft palate. However, all of these anomalies could result in the same disorder of VPI.

32.2.1.1 Overt Clefts

Overt clefts of the palate are the most easily recognizable form of palatal anomaly and may range from complete bilateral clefts of the hard and soft palate to a cleft of the posterior aspect of the soft palate (Fig. 32.5). The extent of the cleft varies in both syndromic and nonsyndromic cases. The frequency of

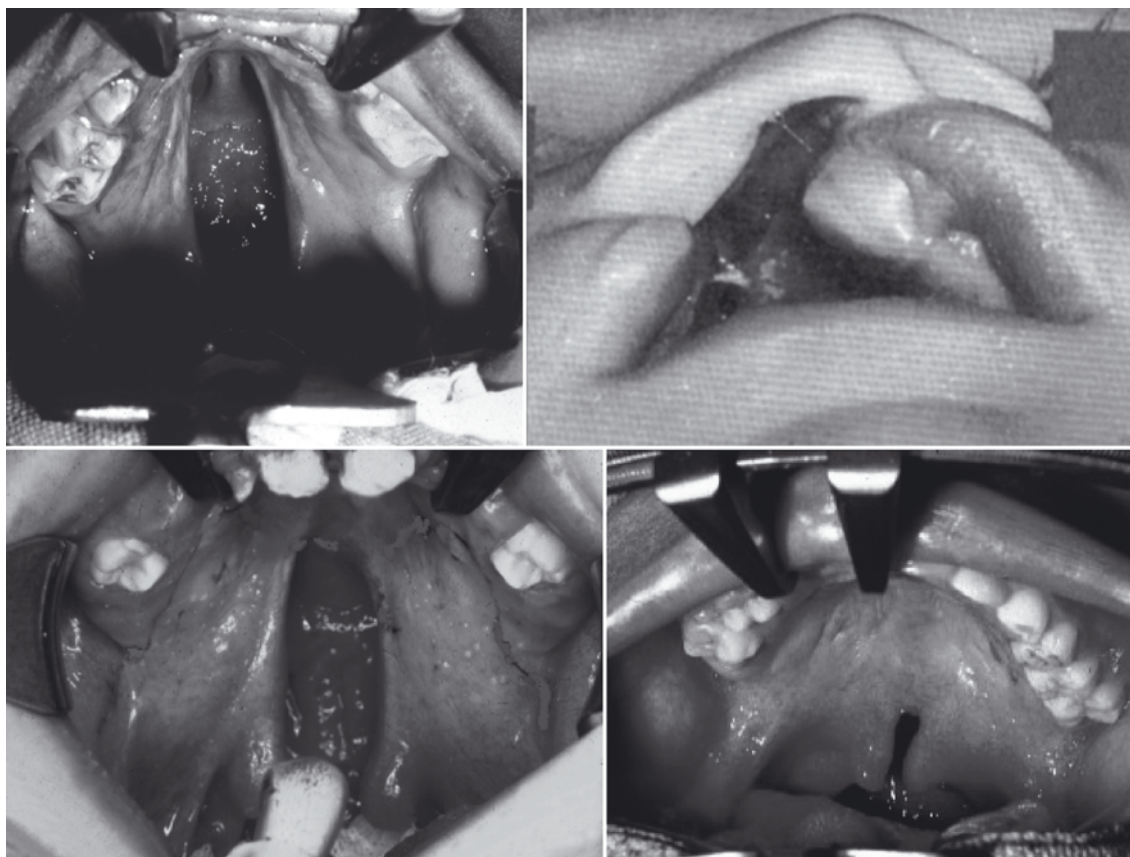


Fig. 32.5. Four different types of overt palatal clefts, including bilateral complete cleft of the hard and soft palate (*top left*), unilateral cleft of the hard and soft palate (*top right*), cleft of the

entire soft palate (*bottom left*), and a cleft of the posterior third of the soft palate

velopharyngeal insufficiency does not really vary according to cleft type, but cleft type does vary according to the primary diagnosis. In nonsyndromic clefts where there are no associated anomalies to affect velopharyngeal closure, the frequency of VPI after repair is probably primarily related to a combination of surgical variables and the amount of muscle deficiency associated with the cleft. However, in syndromic cases that account for more than half of all clefts [7], other syndromic features can contribute to VPI. For example, in velo-cardio-facial syndrome, besides cleft palate, there are three other clinical findings that contribute to VPI, none of which are mutually exclusive. Individuals with VCFS are known to have a high frequency of pharyngeal hypotonia [8], absent or hypoplastic adenoid tissue [9], and platybasia [10] that can increase pharyngeal depth. Therefore, primary cleft repair in VCFS almost always fails to resolve VPI [11].

Surgical repair of the palate is important for only one reason, to provide the patient with the possibility of developing normal speech. Feeding, ear infection, growth...none of these are factors of importance to consider in relation to the need for palatoplasty. Babies with clefts feed quite successfully when the proper technique is used. In fact, because palate repair is not usually completed until approximately 1 year of age, babies must feed over that period of time with a cleft. Middle ear disease is common in babies with clefts, but is related to malformation of the eustachian tube rather than muscular action of the velum [12]. Middle ear disease can also be successfully managed medically or with myringotomy and tubes. Although feeding and hearing can be normal with an unrepaired cleft palate, speech cannot. Achieving acoustically acceptable speech is dependent on normal resonance and the ability to produce pressure consonants orally, both highly dependent on velopharyngeal closure.

Surgical repair of clefts are highly successful when they are accomplished prior to 18 months of age and most surgeons in the USA are timing primary palatoplasty at or before the first birthday in order to intercept the early phases of speech development. It is clear that the majority of children who are born with overt clefts do not have VPI following primary palatoplasty [13]. Although surgical approaches to cleft repair have varied in relation to technique and timing, the majority of experienced surgeons who complete primary palatoplasties prior to 18 months of age achieve between 80% and 90% positive outcomes, defined as an absence of VPI after palatoplasty. Although this leaves a core of patients who will require secondary surgical procedures at a later date to resolve VPI, the overall percentage of individuals with VPI who have repaired clefts is probably quite low in most treatment centers.

The logical conclusion regarding the successful outcome of palate repair is that it is highly dependent on surgical technique. Actually, this has never really been proven and there are many factors involved, including the age at time of surgery, the type of operation, other anatomical variations in the pharynx, muscle tone and developmental factors, and one factor that has received very little attention in the literature, the surgeon and his or her skill level. It is not within the scope of this chapter to discuss each of these issues, but it is important to show that the successful outcome of palate repair is dependent on more than one factor.

Although the palate is an important component of velopharyngeal valving, it is not the only one. The palate, and the velum in particular, is simply one component of the velopharyngeal mechanism and there is quite a bit of variability of the size and shape of the other structures within the pharynx. The height and width of the pharynx varies from person to person just as height, weight, and head circumference does. The size of the adenoid is of particular importance because the adenoid sits in the plane of velopharyngeal closure in young children. In fact, velopharyngeal closure in children until at least 6 years of age is actually velar-adenoidal [9] (Fig. 32.6). Adenoids vary considerably in size and there is some evidence that suggests that adenoid size may be the most important factor for successfully achieving velopharyngeal closure [10]. The larger the adenoid, the less movement is required of the velum. Although the adenoids are important to normal speech production, the tonsils are not and will be considered separately later in this chapter.

Another important factor is the volume of the nasopharynx. An increased volume of the nasopharynx can be caused by an abnormally obtuse angulation of the skull base [11]. Many clinicians who use lateral view radiographs of the head and neck may interpret an increased nasopharyngeal volume as a "deep pharynx." Alternatively, because the palate appears short within the presence of an increased pharyngeal volume, some clinicians may interpret this as a "congenitally short palate." As will be discussed later, structural integrity of the palate cannot be assessed without a direct endoscopic view of the nasal surface of the velum. However, the depth of the nasopharynx can be assessed radiographically using the standardized measurements provided by a lateral cephalograph. Neither procedure (endoscopy or cephalometrics) are individually perfect for describing the anatomy of the pharynx. Rather, the information obtained from each is complementary.

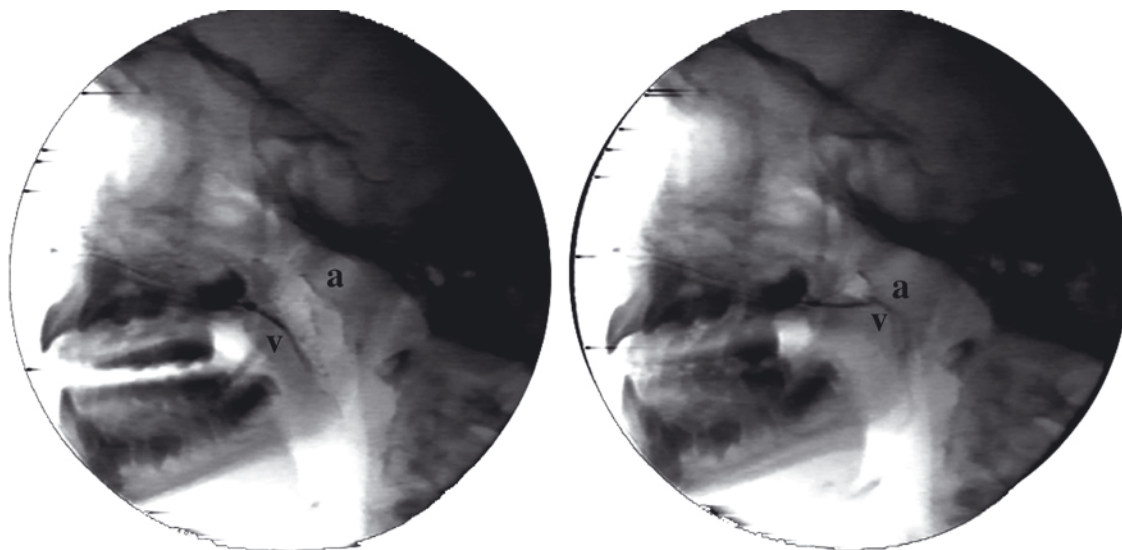


Fig. 32.6. Lateral view videofluoroscopy in a normal 4-year-old child demonstrating that the velum (v) never contacts the posterior pharyngeal wall during speech, but instead approximates to the adenoid (a)

32.2.1.2 Submucous Cleft Palate

Submucous cleft palate has traditionally been defined as the triad of bifid uvula, notching of the posterior border of the hard palate, and a visible diastasis of the musculature in the soft palate referred to as a zona pellucida (Fig. 32.7) [17]. However, although all cases with these clinical findings have submucous cleft palate, not all people with submucous cleft palate have all three of these findings. Some have two of these findings, usually a bifid uvula and hard palate notch, some have only one of these findings, almost always a bifid uvula, and more interestingly, some have none of these findings. Submucous cleft palate is simply a milder or less obvious expression of cleft palate. In this case, the skin, or mucous membrane, is intact but the underlying musculature is not and some muscle may be congenitally absent [18, 19]. Of interest is the fact that although people with submucous cleft palate do have velopharyngeal insufficiency and hypernasal speech, the large majority do not [20]. This is interesting in light of the fact that many surgeons try to resolve speech problems by rearranging the muscles to a more normal configuration in the palate for individuals with submucous cleft palate even though the large majority of people with submucous clefts have normal speech without such a rearrangement. This may indicate that other factors besides muscle integrity play a large or even larger role in normal speech production than simply the degree of palatal normality.

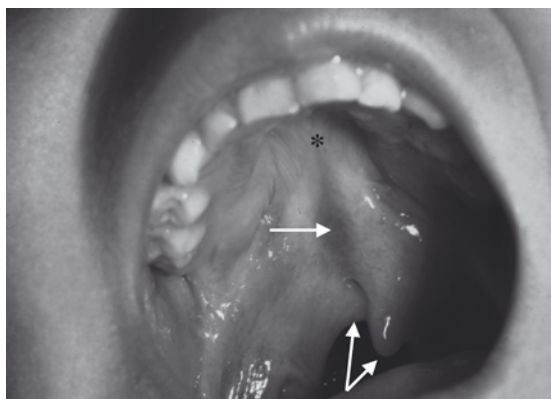


Fig. 32.7. Obvious submucous cleft palate showing a bifid uvula (dual arrow), a notch in the posterior border of the hard palate (*) and zona pellucida (single arrow)

32.2.1.3 Occult Submucous Cleft Palate

In 1975, Kaplan reported an abnormality of the palate he labeled the occult submucous cleft palate [21]. Kaplan's report described four individuals with VPI who had normal appearing palates on oral examination, without evidence of bifid uvula, a zona pellucida, or notching in the hard palate. At surgery for correction of their VPI, he dissected the palate and studied the musculature noting that the muscle fibers were abnormal in position and orientation as might be found

in a cleft palate or submucous cleft palate, hence the label *occult* submucous cleft palate (occult meaning mysterious). He concluded that the only way one could detect this anomaly was by surgical dissection. However, subsequent publications have demonstrated that occult submucous cleft palate can be detected by nasopharyngoscopy [16, 18, 19, 22]. Unfortunately, many clinicians and researchers have not applied this diagnostic approach so that many cases of occult submucous cleft go undetected and the frequency of clefting becomes significantly underestimated. It is interesting to note in retrospect that three of the four patients shown by Kaplan clearly have velo-cardio-facial syndrome, a condition known to have a high frequency of occult submucous cleft palate [23]. It is not known what the frequency of hypernasal speech is in association with occult submucous cleft palate because it is very unlikely that individuals with normal speech will ever have the anomaly detected unless nasopharyngoscopy is performed for some other reason.

32.2.1.4 Other Structural Disorders

Although the adenoid is important to normal speech production, other lymphoid tissue does not play a role and may be detrimental to normal speech production. There are three other masses of lymphoid tissue within the oral and pharyngeal airway: two palatine tonsils and one lingual tonsil. The lingual tonsil sits at the base of the tongue anterior to the vallecula and plays no role whatsoever in speech production. When enlarged, the lingual tonsil can cause the sensation of globus, or a feeling that something is stuck in the bot-

tom of the throat. If very large, a muffled resonance may also result from an enlarged lingual tonsil because it may physically damp the sound waves produced by the vocal cords just millimeters below the base of the tongue. However, the lingual tonsil does not cause hypernasality or hyponasality and is too far away from the velopharyngeal valve to interfere with it. The palatine tonsils, however, may alter nasal resonance patterns as has been documented in the medical literature on several occasions [24–26].

The palatine tonsils are situated in the posterior oral cavity with their attachment in the faucial arch in between the anterior and posterior tonsillar pillar. Assessment of tonsillar size is often relegated to oral assessment with a flashlight and tongue blade. It is typical for clinicians to use a rating scale is used from 0, 1+, 2+, 3+, and 4+ based on the degree of medial excursion of the tonsils towards the midline of the posterior oral cavity with 0 implying that the tonsils are not visible; 1+ referring to tonsils fully contained within the faucial pillars; 2+ meaning the tonsils extrude medially beyond the pillars, but less than halfway to midline; 3+ indicating that the tonsils extend more than half of the way to midline; and 4+ meaning they are touching in the midline (kissing tonsils). The problem with this scale is that no one has told the tonsils that if they get big that they must grow medially. In fact, in many cases they grow back into the pharyngeal airway and may grow down into the hypopharynx or up into the nasopharynx (Fig. 32.8). If the tonsils extrude into the pharynx, they may interfere with velar elevation or lateral pharyngeal wall motion resulting in VPI [24, 25]. In patients with cleft palate, this may present as a particularly difficult diagnostic dilemma and potentially dangerous circum-

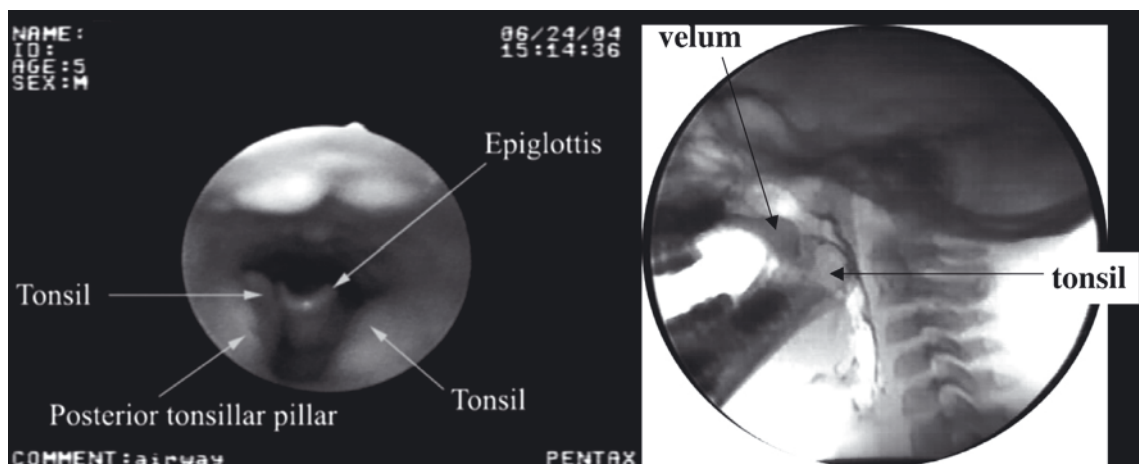


Fig. 32.8. Hypertrophic tonsils seen endoscopically and on lateral view radiographs showing that they intrude posteriorly into the pharyngeal airway behind the faucial pillars

stance. Because VPI is relatively common in children with cleft palate, the presence of hypernasality or audible nasal air escape during speech may lead clinicians to conclude that the problem is an anatomic problem of the palate and therefore a recommendation for pharyngeal flap or other surgery may be made. However, if the tonsils are large and contributing or causing the problem, performing surgery in the pharynx may be unnecessary, but even more important, may prompt the development of obstructive sleep apnea. Tonsils have been shown to be a primary factor in inducing obstructive apnea following pharyngeal flap surgery [27] because they sit beneath the flap and narrow the oropharyngeal airway while the flap compromises the nasal airway. At our Center, we always perform tonsillectomy prior to pharyngeal flap in order to avoid obstructive sleep apnea as a complication and this has proven to be very effective [27]. In some cases, tonsillectomy may resolve the VPI if the tonsils were the primary cause for limiting movement in the velopharyngeal valve [24].

32.3 Neurogenic and Myopathic Disorders of Velopharyngeal Function

VPI is a common disorder among individuals with disorders of the central nervous system or who have primary myopathies. Although the frequency of VPI is high, the true prevalence is not known because many of these individuals have other severe disorders of speech and the VPI is simply a component of a broader spectrum of speech abnormality that may be very severe. In such cases, resolving VPI alone will do little or nothing to improve the intelligibility of speech and therefore such individuals are not often referred for diagnosis or treatment. Neurogenic disorders that might frequently result in VPI are strokes and other cerebrovascular accidents, open and closed head trauma, cerebral palsy resulting from neonatal hypoxia or brain malformation, infections such as encephalitis, and neurodegenerative diseases, such as myasthenia gravis. Primary myopathies that have VPI as a common finding include (but are not limited to) Steinert syndrome (myotonic dystrophy), facioscapulohumeral muscular dystrophy, nemaline myopathy, and oculopharyngeal muscular dystrophy. The management in myopathies is very different than in cleft palate or other structural anomalies of the head and neck. Surgical intervention is often problematic in myopathies because of respiratory concerns, and even the operation itself may be compromised by severe adverse anesthesia reactions, particularly the development of malignant hyperpyrexia which can be a fatal complication [28]. Therefore, the diagnosis of myopathic dis-

orders and the recognition of hypernasal speech as a potential clinical feature are critical for proper clinical management.

32.4 Evaluation of Velopharyngeal Insufficiency

Two points established in the previous pages point out the need for detailed and comprehensive evaluation of VPI. First, velopharyngeal valving is highly variable from person to person, both in normals and in those with velopharyngeal insufficiency. Therefore, assumptions cannot be made about the nature of velopharyngeal valving based on the sound of speech or preconceived notions about how the mechanism works for the “average” individual. Second, VPI occurs in many different conditions and the reason for the VPI may not always be obvious, including structural anomalies that cannot be seen on oral clinical examination. The diagnosis of more obscure yet common conditions such as occult submucous cleft palate, tonsillar hypertrophy, or myopathies is highly dependent on a precise diagnostic approach.

In general, procedures used to assess velopharyngeal insufficiency can be divided into two broad categories: direct and indirect. Direct procedures can be defined as those that allow direct visualization of the velopharyngeal valve during the unimpeded flow of speech typical for the individual while not interfering with the movements of the valve. Indirect procedures can be defined simply as everything else. Indirect procedures typically measure some type of artifact or by-product of velopharyngeal valving rather than assessing the act itself. Indirect procedures consist of listener judgment of resonance and nasal airflow, instrumental techniques for measuring nasal resonance, and techniques for measuring nasal airflow or pressure. Direct procedures include both radiographic and endoscopic techniques, although ultrasound and real time CT and MRI have all been proposed to study VPI. However, only fluoroscopic and endoscopic procedures have stood the test of time and remained.

32.5 Indirect Assessments of VPI

Every diagnostic process starts with an assessment of the way a patient sounds to the human ear. Parents may perceive something is wrong with the way the child’s speech sounds or, if the child has a repaired cleft, a clinician schedules periodic assessments to determine if speech will develop normally. At some point, however, someone has to perceive abnormal hypernasality or nasal air escape during speech. The listener must determine if resonance is normal, hy-

pernasal, or hyponasal. Occasionally there may be mixed hyper-/hyponasality if there is VPI in association with nasal blockage at some location anterior to the velopharyngeal valve. In addition to resonance, the listener must determine if there is an abnormal escape of air during speech, and if so, on what sounds and how often it is occurring. If the speech pattern is deemed to be abnormal, then the necessity for treatment is determined. In other words, perceptual assessment becomes the entry point for further evaluation and is therefore the most critical portion of the assessment.

Once a patient's speech has been determined to show the effects of VPI, then the clinician must choose what to do next. The clinician may choose to confirm nasal air escape. This can be done in a number of ways from very simple to very complex. Simple procedures would include the use of a nasal mirror, a listening tube or stethoscope, or devices that might detect nasal air escape such as the SeeScope. All of these devices are used to confirm the presence of abnormal nasal air escape during the utterance of normally nonnasal sounds, words, or phrases. They are "yes vs. no" devices rather than quantitative instruments. Although sensitive to nasal air escape, they are not specific as to the source of the air escape. In individuals with clefts, the air may be leaking from a fistula even if there is no VPI, thus resulting in a false positive. False negatives are common. Nasal congestion or a deviated septum will block air escape from the nostrils even if VPI is present, resulting in a false negative. Therefore, procedures for detecting nasal air escape may be decent "quick and dirty" techniques for identifying nasal air escape, but they are not really diagnostic for VPI in many cases. Perhaps the easiest procedure for a "quick and dirty" technique is to use nostril occlusion during normally nonnasal speech. Occluding the nostrils during a normally nonnasal speech sample should result in no change of resonance if velopharyngeal closure is being achieved. If there is VPI, then pinching the nostrils shut will result in a cul de sac nasal resonance that will cause an immediate change in resonance pattern. If nasal air escape is audible during spontaneous speech, occluding the nostrils will eliminate the nasal turbulence. Although this procedure is not quantitative in any way, it is a simple procedure for making a quick determination of the potential presence of VPI.

There are several devices that have been designed as quantitative. These include manometers and rhinometers that measure pressure from the mouth or nose, but these devices have been proven to be irrelevant in relation to speech production. Two procedures have garnered some attention from their use in research and some clinical settings: the nasometer and pressure-flow instrument. The nasometer and pres-

sure-flow device both apply numbers to nasal air escape during speech based on resonance (nasometer) or the pressure of air coming from the nose and mouth (pressure-flow) during speech. The nasometer uses microphones to differentiate resonance between the nasal and oral cavities while the pressure-flow instrumentation utilizes a mathematical computation, the theoretic hydraulic principal, to calculate an orifice size in mm^2 . Although these devices may be used for biofeedback purposes, we do not use them at our Center because we do not equate a number with "objective" data. Albert Einstein was reputed to have a sign hanging in his office at Princeton that said, "Not everything that counts can be counted, and not everything that can be counted counts." Further, relevant to the hydraulic principal calculation, Einstein also said, "As far as the laws of mathematics refer to reality, they are not certain; and as far as they are certain, they do not refer to reality" and "If the facts don't fit the theory, change the facts." In other words, validity of a measure has more to do with its relevance than anything else. The nasometer is a device for detecting resonance patterns, but the ultimate tool for this is the examiner's ear. It is the perception of normal versus abnormal that is important. If the ear perceives speech as normal, it matters not what a machine assigns as a numerical value. The second and third quotes from Einstein reinforce a primary problem with pressure-flow instrumentation. The entire principle behind pressure flow is that the opening detected in the velopharyngeal valve is measured in mm^2 . As was emphasized earlier in this chapter and as is shown in Fig. 32.1, the velopharyngeal valve is a three dimensional structure, not two dimensional. A three dimensional object needs to be measured in mm^3 , not mm^2 . This misinterpretation of velopharyngeal valving that serves as the basis for pressure-flow studies renders its calculations as irrelevant.

32.6 Direct Assessments

32.6.1 Multiview Videofluoroscopy

It is interesting to note that the two gold standards for direct assessment of velopharyngeal valving were both initially described in the same year. Multiview videofluoroscopy and nasopharyngoscopy were both described initially in 1969, and although there have been improvements in instrumentation and minor changes in technique, the procedures have remained relatively unchanged since that time. Both procedures are used widely and were recognized by an international working group as being critical to the proper assessment of VPI [29].

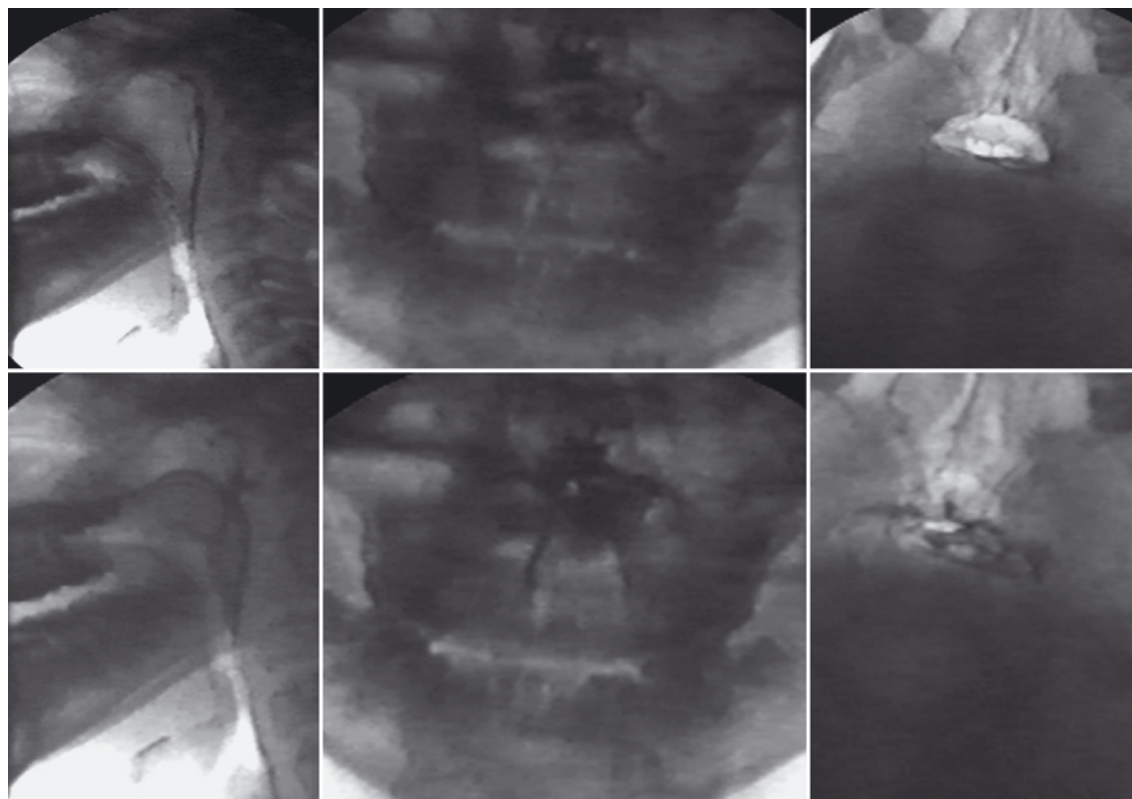


Fig. 32.9. Lateral, frontal, and base radiographic views at rest and during speech in an individual with a repaired cleft and hypernasal speech

In 1969 and 1970, Skolnick described the use three radiographic views, the lateral (midsagittal), frontal (P-A), and base (axial) views using barium contrast to coat the pharyngeal soft tissues [30, 31] (Fig. 32.9). Although the views were not performed simultaneously, the use of a standardized speech sample serially provided a three-dimensional assessment of the pharynx that accounts for width (frontal and base views), depth (lateral and base views), and height (lateral and frontal views). In addition, all of the oral articulators can also be seen simultaneously. Since the early descriptions of this technique, several other radiographic views have been added, including oblique views and Townes view [32]. According to the International Working Group, the two views essential to assessment are the lateral and frontal [29] and other views may be added if they provide useful additional information. A standardized speech sample is used to limit the duration of radiation exposure while allowing an observation of all components of speech including nonnasal sounds, nasal sounds, syllable repetitions, a sustained fricative, nasal to nonnasal transitions, and the natu-

ral flow of spontaneous speech. We currently use the following sample that meets these criteria:

- Ma-ma-ma
- Pa-pa-pa
- Ta-ta-ta
- Ka-ka-ka
- /ssssss/ (sustained /s/)
- Suzy
- Suzy sees Sally
- Stop the bus
- Catch a fish
- Jerry's slippers
- 1-2-3-4-5-6-7-8-9-10 (counting to 10)

The speech sample is rehearsed with the patient just prior to the procedure to be certain that it is repeated correctly and in a conversational manner. Repeating this sample for three radiographic views limits the procedure to a minute or less, thus keeping radiation exposure to a minimum. Because the neck is irradiated during this procedure and a child's thyroid gland is sensitive to radiation, we prefer to limit videofluo-



Fig. 32.10. Arrangement of equipment for videonasopharyngoscopy. Two television monitors are used to allow both the patient and the examiner to see the examination simultaneously.

roscopy to a one-time evaluation, usually just prior to secondary surgical intervention.

Immediately prior to the procedure, approximately 1 ml of barium sulfate suspension is instilled in each nostril using a dropper or pipette. This is done with the patient supine on the fluoroscopic table. This allows coating of the velum, lateral pharyngeal walls, and posterior pharyngeal wall. Without barium contrast, the lateral pharyngeal walls cannot be seen in frontal view, and the base or Townes view will not provide an en face view of the actual velopharyngeal valve. In addition, even in lateral view the barium contrast is essential for defining small gaps. If there is no contrast used, as soft tissues converge they blur together and a false negative for VPI is possible.

32.6.2 Nasopharyngoscopy

Nasopharyngoscopy for the assessment of the velopharyngeal valve was described almost simultaneously with the advent of multiview videofluoroscopy. Pigott described the use of both flexible and rigid instruments in 1969 [33, 34]. Although Pigott continued to advocate for the use of rigid endoscopes in the years that followed [35], the development of smaller caliber flexible instruments clearly won support for fiberoptic endoscopy because of the ability to examine even very young children without significant pain or discomfort [36–38]. Today, fiberoptic endoscopes have external diameters well under 3 mm, many in the 2 mm range, therefore allowing easy access to the upper airway (Fig. 32.10). The International Working Group recommended flexible fiberoptic

This helps to fix the attention of the patient and allows the examiner to explain the observations while the examination is taking place

nasopharyngoscopy and further recommended that the examination should include assessment of the entire vocal tract from the tip of the nose to the larynx [29]. It was also recommended that all examinations be videotaped for future review [29]. It has been shown that group review of studies is the most reliable and valid method for coming to diagnostic and treatment decisions because it allows for discussion of interpretations of the procedure [39].

The procedure is best done using topical anesthesia, especially in young children, in order to avoid discomfort so that an adequate speech sample is obtained. My preference is to use topical tetracaine hydrochloride 2% aqueous solution (Pontocaine) inserted into the nose in cotton packing rather than a spray. Using a pair of bayonet forceps, the cotton packing can be passed deeply into the nose to numb the turbinates and septum which are the sensitive portions of the nose. The packing is left in place for several minutes, then removed and the endoscope inserted. Using packing, the anesthetic is kept against the nasal mucosa for several minutes rather than a brief spray which really has minimal effect. In addition, the spray may reach deeper into the vocal tract and be inhaled whereas the packing keeps the effect limited to the nasal mucosa. Once the endoscope is passed into the pharynx, it should not be held stationary throughout the examination. The advantage of a flexible instrument is its flexibility that allows movement so that both sides of the pharynx can be seen and it can be passed into the nasopharynx, oropharynx, and hypopharynx. The larynx, tonsils, and tongue can be seen in this manner.

The same speech sample used for videofluoroscopy is utilized for nasopharyngoscopy, as well, but the sample does not need to be limited to those sounds and phrases. Because there is no ionizing radiation involved, there is no need to limit the procedure to a minute or less. If phonemic variation is seen, the examiner is free to have the patient say more sounds and phrases than for fluoroscopy.

32.7 Which Direct Visualization Procedure to Use?

Many clinicians utilize nasopharyngoscopy exclusively as an assessment of VPI because they think that the information provided is the equivalent of videofluoroscopic procedures. However, it has been reported that there is an approximate 30% discrepancy between findings using multiview videofluoroscopy and nasopharyngoscopy primarily in the rating of lateral pharyngeal wall motion [32]. Therefore, both procedures are essential for a complete assessment of VPI [29]. Each procedure has its own advantages and disadvantages as follows:

32.7.1 Multiview Videofluoroscopy Advantages

1. Provides a direct physiologic view of all components of velopharyngeal valving in all dimensions (width, depth, and height).
2. The entire vocal tract can be seen at one time.
3. All articulators are seen simultaneously.
4. The forward floor of speech is not impeded.
5. Excellent compliance even in very young children.
6. The procedure is painless.

32.7.2 Multiview Videofluoroscopy Disadvantages

1. It is not a direct anatomic view, relying on radiographic shadows for information.
2. Ionizing radiation is used.
3. As a result of ionizing radiation, there is a time limit to the procedure.
4. Interpretation may be difficult for those inexperienced with the technique.

32.7.3 Nasopharyngoscopy Advantages

1. Provides a direct anatomic view usually appreciated by surgeons.

2. The entire vocal tract can be seen, but not simultaneously.
3. The normal flow of speech production is not impeded.
4. There is no time limit to the procedure so speech can be manipulated in more detail.
5. There is no ionizing radiation.
6. The procedure should be painless when done properly.

32.7.4 Nasopharyngoscopy Disadvantages

1. Anatomic structures moving in front of the endoscope may block the view of physiological movements.
2. A lack of compliance occurs in some cases, particularly in young children.
3. The oral articulators cannot be seen.
4. The entire vocal tract cannot be seen at the same time.
5. The procedure can be painful in some cases or cause bleeding if not done properly.

32.8 Reporting of the Results

The International Working Group also published guidelines for the standardized reporting of the observations from endoscopies and fluoroscopies [29]. A ratio scale was developed that would assess the amount of movement of a structure (the velum, right lateral pharyngeal wall, left lateral pharyngeal wall, and posterior pharyngeal wall) towards its opposing structure. This approach was an attempt to provide a method that would communicate the degree of movement, not the actual distances moved, that would be more standardized than qualitative measures such as “poor,” “fair,” “good,” or “excellent.” We have been utilizing this system at our Center with a great deal of success in terms of the ability to communicate between clinicians, especially those from diverse fields of practice, such as surgeons, speech pathologists, and otolaryngologists.

32.9 The Importance of It All

There is a common “Murphyism” (one of the humorous attempts at expanding Murphy’s Laws) that goes something like this, “Before ordering a diagnostic test, determine what you will do if the result is positive or the result is negative. If the answer is the same for both, don’t do the test.” In other words, the value of collecting such elegant diagnostic information is to

have it guide treatment. This has been amply demonstrated in a number of studies that have documented the efficacy of guiding surgical management based on diagnostic information from multiview videofluoroscopy and nasopharyngoscopy [38, 40–42]. Stated another way, it has become obvious that there is not one single operation that can be successful in all cases. A velopharyngeal gap during speech is a hole that needs to be located in three-dimensional space and its size relative to the total size of the pharynx must be confirmed in order to know how to occlude it. The listener's ear becomes the final judge of success, just as it was the entry point into the diagnostic protocol. With the combination of direct visualization procedures and an understanding of how to structurally solve the problem, the elimination of VPI should be achieved nearly every time.

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33 The Nasal Airway in Breathing and Speech

DONALD W. WARREN, AMELIA F. DRAKE

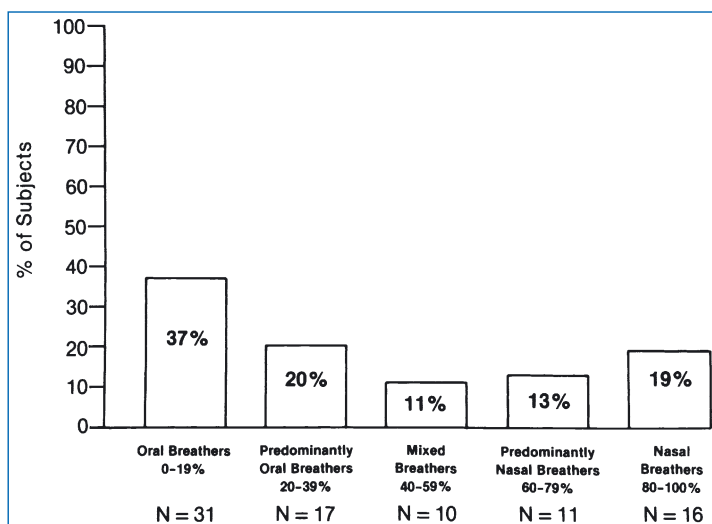
Clefts of the lip and palate frequently produce significant nasal deformities, such as deviated septum, vomerine spurs, and atresia of the nostrils, as well as maxillary growth deficits that alter the nasal floor [1–3]. These abnormalities tend to reduce the size of the nasal airway.

Drettner [1] reported that septal deformities, atresia of the nostrils, and turbinate hypertrophy diminish airway size and increase airway resistance. Interestingly, in studies previously performed in our laboratory [2], otolaryngologic examinations revealed that about 60% of the “normal” subjects demonstrated clinical evidence of septal defects, vomerine spurs, or turbinate hypertrophy in spite of the fact that their nasal airways were judged to be normal. Approximately 70% of the subjects with cleft lip and palate also presented with similar nasal defects, although these abnormalities were usually more severe. Those with cleft lip and palate had a greater inci-

dence of nasal deformities than those with cleft palate alone. We determined that nasal resistance in individuals with cleft palate is about 20%–30% higher than in the normal population [2, 3].

There is evidence that airway deficiency in unilateral cleft lip and/or palate is present in utero. Siegel et al. [4] observed that the fetal septum appeared enlarged and distorted and was flanked laterally by reduced nasal airway passages at 17 weeks of gestation. They suggest that reduced nasal size may be a function of both nasal capsule deficiency and nasal septum hypertrophy. Additionally, histochemical abnormalities in facial muscles of unrepaired clefts, including mitochondrial abnormalities and muscle fiber hypoplasia, suggesting a myopathy or delayed maturation, have also been demonstrated [5]. These abnormalities may affect the nasal musculature as well. In repaired clefts, abnormal muscle bundles and connective tissue have also been identified at some

Fig. 33.1. Percentages and number of individuals according to respiratory classification



distance from the cleft margin [6]. Their appearance is consistent with surgical denervation of seventh nerve muscle groups in the perialar region. We recently reported that the nasal airway of an adult with a repaired cleft lip and/or palate generally is about 25% smaller than that of a normal adult [3]. The normal adult nose is about 0.62 cm² in the smallest cross-sectional area, while the nose of an adult subject with cleft is about 0.43 cm² [2, 3, 7, 8]. Evidence that the cleft airway is narrow implies that the airway is impaired and mouth-breathing is common. Indeed, a recent study by Hairfield et al. [9] confirms this. Approximately 70% of the subjects with cleft lip and/or palate evaluated were either oral, predominantly oral, or mixed oral-nasal breathers (Fig. 33.1). Although a high prevalence of mouth-breathing was expected in the cleft population, the extent and severity observed were not anticipated. Published data on the prevalence of mouth-breathing in the normal population indicate that the percentage is approximately 15% [10–12]. Our continuing studies suggest that about 30% of the normal population breathes through the oral mode to some extent.

There is only one report at variance with previous findings. Sandham and Solow [13] did not find a difference in nasal airway resistance between subjects with clefts and a normal control group. The authors suggested that the difference between their results and others may be attributable to their using a decongestant prior to measurements.

33.1 Children

In children, the type of cleft appears to affect nasal airway size, but not necessarily the breathing mode [14]. Table 33.1 presents data on nasal airway size during inspiration and expiration according to type of cleft. Normative data also are shown for comparison. Differences among cleft types are readily apparent. The group with bilateral cleft lip and palate and the group with unilateral cleft lip demonstrate nasal cross-sectional areas that are significantly larger than the other cleft types. Indeed, the group with bilateral cleft lip

and palate has an area slightly larger than the normal group.

Table 33.2 lists data on percent of nasal breathing by cleft type. The criteria used to categorize percent of nasal breathing are as follows. Individuals who demonstrated 80% nasal breathing or above were classified as nasal breathers. Individuals who scored between 60% and 80% were classified as predominantly nasal breathers. Those falling in the 40%–60% subgroup were considered mixed oral-nasal breathers. The 20%–40% subgroup comprised the predominantly oral breathers, and those in the 0%–20% subgroup were considered to be oral breathers. These are arbitrary classifications, but appear to be reasonable categorizations [15, 16].

It is especially noteworthy that 75% of the individuals in the bilateral cleft lip and palate group were more oral than nasal breathers. The percentage is similar to that observed among the group with unilateral cleft lip and palate, even though the latter group had significantly smaller airways. Only the group with unilateral cleft lip only had a greater number of nasal than oral breathers. The data from Table 33.3 reveal that the means for most cleft types fall in the mixed oral-nasal breathing category. Only the means for the group with unilateral cleft lip and the normal group fell in the predominantly nasal breathing category.

Although a reduced nasal air space was expected for subjects with cleft lip and palate, the difference between the subjects with unilateral and bilateral cleft lip and palate was not anticipated. Previously, only differences between cleft lip and palate and cleft palate have been noted. Drettner [1] reported that individuals with cleft lip and palate had a narrower nasal airspace than those with cleft palate alone. Sandham and Solow [13] although not directly comparing the two groups, did show a slight, but not statistically significant, difference between these groups.

The nasal valve is usually the site of the smallest cross-section of the nose and the region of greatest air flow resistance [17–19]. However, in the cleft population, other areas may be involved as well. Otolaryngologic examination revealed that septal deformities occurred most frequently. However, stenosed nasal

Table 33.1. Nasal airway size by type of cleft

	Nasal Airway Size in cm ² (Mean + SD)					
	UCL	UCLP	BCLP	CHP	CSP	Normal
Inspiration	0.37 ± 0.22	0.21 ± 0.13	0.40 ± 0.15	0.26 ± 0.14	0.26 ± 0.05	0.36 ± 0.18
Expiration	0.36 ± 0.20	0.22 ± 0.12	0.41 ± 0.14	0.27 ± 0.15	0.23 ± 0.05	0.34 ± 0.17

UCL, unilateral cleft lip; UCLP, unilateral cleft lip and palate; BCLP, bilateral cleft lip and palate; CHP, cleft of hard palate; CSP, cleft of soft palate.

Table 33.2. Subjects categorized by mode of breathing and cleft type

Group	Mode of breathing (number of subjects)				
	Oral (0%–19% nasal)	Predominantly oral (20%–39% nasal)	Mixed oral-nasal (40%–59% nasal)	Predominantly nasal (60%–79% nasal)	Nasal (≥80% nasal)
UCL (n = 9)	1	0	3	2	3
UCLP (n = 16)	6	3	3	1	3
BCLP (n = 8)	3	1	2	0	2
CHP (n = 8)	2	1	2	1	2
CSP (n = 9)					
Normal (n = 95)	3	1	1	1	3
Normal (n = 95)	11	16	10	12	46

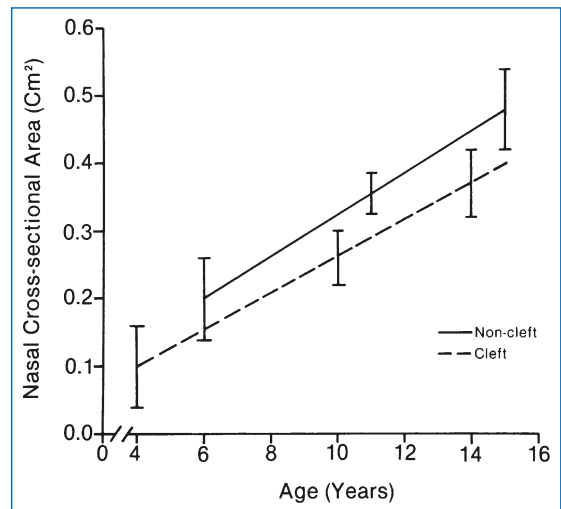
UCL, unilateral cleft lip; UCLP, unilateral cleft lip and palate; BCLP, bilateral cleft lip and palate; CHP, cleft of hard palate; CSP, cleft of soft palate.

Table 33.3. Percentage of nasal breathing in each group

Percentage of nasal breathing (mean ± SD)					
UCL	UCLP	BCLP	CHP	CSP	Normal
66 ± 28	41 ± 37	43 ± 32	50 ± 32	53 ± 14	69 ± 35

UCL, unilateral cleft lip; UCLP, unilateral cleft lip and palate; BCLP, bilateral cleft lip and palate; CHP, cleft of hard palate; CSP, cleft of soft palate.

apertures, thickened nasal mucosa, and hypertrophied turbinates also were observed. Similarly, the collapsed nasal ala associated with the typical cleft lip-nasal deformity was frequently observed, especially in the group with unilateral cleft lip and palate. The validity of equating subjective clinical observations of nasal deformities with objective measurements is questionable. However, some comment is appropriate. There is evidence that conditions or procedures that alter the shape or function of the nasal valve also affect nasal cross-sectional area. As noted earlier, histologic abnormalities in the perialar musculature suggestive of surgical denervation/reinnervation have been demonstrated and undoubtedly adversely affect the nasal valve on which they insert [6]. Hall et al. [20] and Guenther et al. [21] demonstrated that superior repositioning of the maxilla increases nasal area, presumably by changing the shape of the nasal valve. Cosmetic procedures that inappropriately scar or cause collapse of upper lateral cartilage also narrow the airway [22]. In fact, any morphologic changes in the critical nasal valve region may influence mode of breathing. Individuals with bilateral cleft lip and palate usually undergo surgery that lengthens the columella. Presumably, this procedure should have a positive effect on alar shape and widen or stent the nasal valve. It is quite possible that the difference in airway

**Fig. 33.2.** The relationship between age and cross-sectional area

sizes noted between the group with bilateral cleft lip and palate and the group with unilateral cleft lip and palate stems from this surgery. Although the difference was not statistically significant, it is interesting to note that, even groups without lip involvement, namely, the groups with cleft of the hard and soft palate and clefts of the soft palate, had somewhat smaller airways than normal.

Another factor that affects nasal airway size is age. Figure 33.2 demonstrates that nasal cross-sectional area increases with age in the cleft, as well as the non-cleft, populations. However, the 25%–30% difference in airway size apparently remains over time despite the increase in nasal airway size. Similarly, for the noncleft population, breathing becomes more nasal with increased age. However, the cleft group consis-

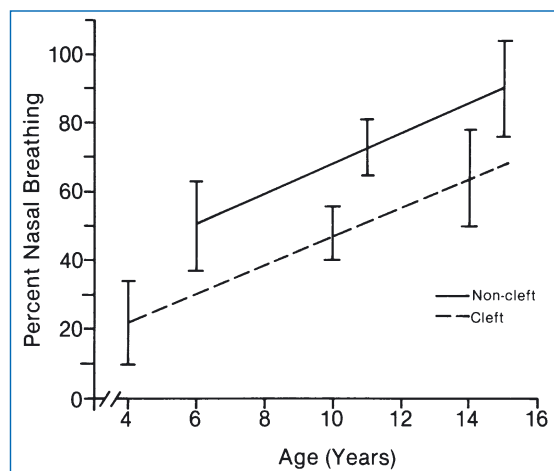


Fig. 33.3. The relationship between age and percent nasal breathing

tently remains behind the noncleft group (Fig. 33.3). These findings pose interesting clinical questions. Specifically, all palate-involved groups, including the group with bilateral cleft lip and palate, were more oral than nasal breathers. Based on airway size, the group with bilateral cleft lip and palate should have had a higher mean percentage of nasal breathing. In fact, a percentage of nasal breathing similar to that observed in the group with unilateral cleft lip would be expected. Although it is only conjecture, we suggest that distortion of the nasal valve and alar collapse, so frequently associated with bilateral cleft lip and palate prior to initial repair and columellar lengthening, resulted in high nasal resistance.

Because there is open communication between the nose and mouth in newborns with clefts, a normal nasal breathing pattern is not established. Additionally, endoscopic studies of some of our bilateral cleft lip and palate patients indicate that, prior to palatal repair, the tongue is often placed in the nose during breathing. These conditions may produce a persistent pattern of mouth-breathing that remains even after surgical repair. Hairfield et al. [9] also noted persistent mouth-breathing. That is, mouth-breathing patterns in individuals with clefts tend to become habitual over time.

These findings are at variance with what occurs in the noncleft population. Linder-Aronson [23, 24] reported that children with adenoidal obstruction return to a nasal breathing mode after adenoidectomy. Perhaps the difference in results arises from the fact that the children with adenoid obstruction were nasal breathers at birth, whereas the cleft subjects were not.

If obligatory mouth-breathing becomes habitual mouth-breathing in this population and the precipi-

tating factor is early airway impairment, a change in surgical approach might be considered. Clinicians might consider the possible advantages of early surgical intervention to reduce airway impairment. Elimination of nasal deformities, especially septal defects that interfere with nasal valve function, might prove beneficial. This must be balanced, of course, with the need to provide treatment that does not interfere with nasomaxillary growth [25]. Additionally, alteration of surgical techniques to prevent denervation of perialar musculature appears warranted. Presurgical molding may provide an improved airway by avoiding scarring in the nasal valve area. Early results suggest that the procedure also minimizes the extent of primary surgery [26].

33.2 Adults

Type of cleft does not appear to have the same effect in adults. As Table 33.4 indicates, there is no difference in nasal cross-sectional area between the unilateral cleft lip and palate and the bilateral cleft lip and palate. Although the mean unilateral cleft lip and palate nasal area almost doubled (i.e., 0.22 cm² in children at a mean age of 9.7 years versus 0.41 cm² in adults), less change occurred with age in the bilateral cleft lip and palate group. In fact, during inspiration, the bilateral cleft lip and palate nose tends to collapse with the negative inspiratory pressures. This apparent lack of growth in the bilateral cleft lip and palate group is a concern. The columellar lengthening procedure apparently provides an adequate nasal airway for most children, but the data for adults suggest that this cosmetic surgical procedure may, in fact, hinder further growth in the critical area of the nasal valve, especially if muscle disinsertion or denervation occurs. We are presently assessing this disturbing finding. Similarly, the fact that the nasal area is 0.11 cm² smaller during inspiration indicates that the nasal valve is somewhat flaccid in this group of patients, a finding that has implications for speech as well as breathing.

Table 33.4 also lists the percentage of individuals with impaired nasal airways by type of cleft. The data also confirm [7] the presence of impairment (i.e., nasal cross-sectional area less than 0.40 cm²) in a significant proportion (60%) of the adult cleft population. Studies of normal and nasally impaired adults [27, 28], modeling studies [3], and extrapolation of data from nasal resistance studies [29–31] suggest that airway impairment in adults occurs when the smallest cross-sectional area anywhere in the nose is less than 0.40 cm². Areas greater than 0.40 cm² during rest breathing usually meet respiratory requirements without physical constraints imposed by the relation-

Table 33.4. Comparison of areas and percentages with impairment between subject groups

Group	Inspiratory (cm ² + SD)	Expiratory (cm ² + SD)	Nasal Area <0.40 cm ² (%)
Normal	0.63 ± 0.17	0.56 ± 0.14	20
UCLP	0.41 ± 0.14	0.41 ± 0.13	45
BCLP	0.34 ± 0.23	0.45 ± 0.29	67
CP	0.35 ± 0.17	0.32 ± 0.16	67
U/BCL	0.40 ± 0.16	0.44 ± 0.13	60

UCLP, unilateral cleft lip and palate; BCLP, bilateral cleft lip and palate; CP, cleft palate; U/BCL, unilateral and/or bilateral cleft lip.

ship of area to airflow rate [28]. Other studies indicate that adults with cross-sectional areas less than 0.40 cm² mouth-breathe to some extent [16, 28, 29, 32].

33.3 Effects of Secondary Procedures on the Nasal Airway

Secondary procedures for velopharyngeal inadequacy, such as the pharyngeal flap, alter the nasopharyngeal airspace, increase nasal airway resistance [33], decrease airway size [7], and increase the prevalence of mouth-breathing [9]. Figure 33.4 illustrates that patients with flaps tend to be more impaired than those without flaps. However, the placement of a flap does

not necessarily mean that an individual will notice or complain of increased airway resistance. As Fig. 33.5 demonstrates, 78% of subjects with flaps are predominantly oral breathers. On the other hand, as Fig. 33.1 shows, 68% of the patients without flaps are substantial oral breathers. Thus, the addition of a flap really does not change breathing behaviors in most individuals. However, the effect might be significantly different in an individual whose airway and breathing mode were fairly normal prior to the flap, but impaired after the flap. Under such circumstances, the individual would have to switch to some oral breathing and probably notice that breathing was more difficult. In some instances, pharyngeal flaps may even result in sleep apnea, especially in children [33–35]. Similarly, when adenoid tissue hypertrophies around the flap or when enlarged tonsils are present, the ports may become obstructed. The removal of both tonsils and adenoid tissue prior to pharyngeal flap surgery should be considered in light of possible life-threatening consequences [36].

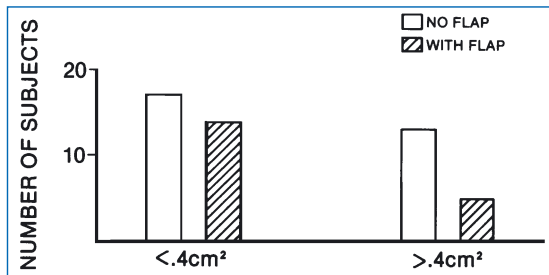
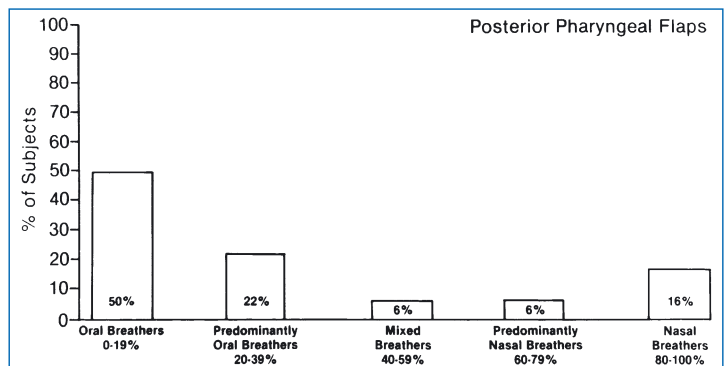


Fig. 33.4. The data are presented in two categories, subjects whose nasal size was <0.4 cm² and those nasal size was >0.4 cm². In adults, an area less than 0.4 cm² is considered impaired

33.4 Speech Aid Prostheses and the Nasal Airway

Prosthodontists are frequently called on to make speech aid prostheses for individuals with velopharyngeal inadequacy. The prosthesis obturates a considerable portion of the nasopharyngeal airspace,

Fig. 33.5. Findings for subjects who had received posterior pharyngeal flaps



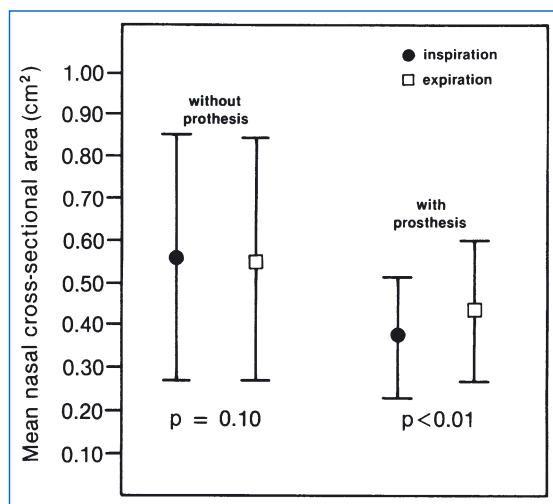


Fig. 33.6. Comparison of mean nasal cross-sectional areas for inspiratory and expiratory phases of respiration with and without speech aid prosthesis present in oral cavity

creating a constriction within the airway. Minsley et al. [37] assessed the effects of such appliances on nasal cross-sectional area and reported that the nasopharyngeal airway is reduced by approximately 30% when the speech aid is in place. Almost 50% of the adult subjects had an airway during breathing that was less than 0.4 cm^2 with the appliance in place (Fig. 33.6). Additionally, the negative pressures associated with the inspiratory phase of breathing apparently closed the airway even more in most subjects. Thus, some mouth-breathing would be expected in nearly all individuals with a prosthetic speech aid.

33.5 Effects of Maxillary Expansion on the Nasal Airway

Individuals with cleft palate often have maxillary arch constriction that requires further treatment. In some cases, expansion by orthodontic means is sufficient, but in others, surgical expansion may be necessary. Both procedures appear to have a beneficial effect on nasal respiration [20, 38, 39]. Figure 33.7 illustrates the gain in nasal airway size after 1 year of orthodontic expansion. A similar result for surgical expansion is shown (Fig. 33.8). An increase in the range of 45%–55% can be expected, although, in many instances, this increase is still not great enough to ensure a change to nasal breathing.

Both procedures apparently alter the nasal valve area, which in the normal nose presents the smallest nasal cross-sectional area and provides the most significant airflow resistance during breathing [18].

Aerodynamic studies of the nasal airway support this assumption. Hall et al. [20] and Guenther et al. [21] suggest that maxillary repositioning opens the nasal valve, thus reducing nasal resistance. They observed that the external nares change in shape from narrow slits to more ovoid forms postoperatively. Rapid maxillary expansion probably has a similar effect because suture opening is greatest in the anterior palate [40]. A reasonable assumption is that maxillary constriction produces a narrow nasal valve. Although nasal airway resistance may be increased by turbinate hypertrophy, nasal polyps, extremely large adenoids, and a deviated septum, maxillary expansion should have little effect on such factors. Expansion does appear to improve airway patency by increasing alar width and nasal valve size [20, 21, 39, 41].

An interesting additional finding was that nasal breathing also improves after expansion. Walker et al. [42] noted a considerable change toward nasal breathing in most subjects who underwent a maxillary osteotomy. They noted that, although individuals who were oral-nasal breathers for a long period of time, such as in the cleft population, tend to remain oral breathers even with improvement in nasal airway patency, this was not the case in their study. They suggested that maxillomandibular fixation for a period of 6–8 weeks may have encouraged a change to nasal breathing. Faced with high oral airway impedance due to fixation, many may have changed to lower resist-

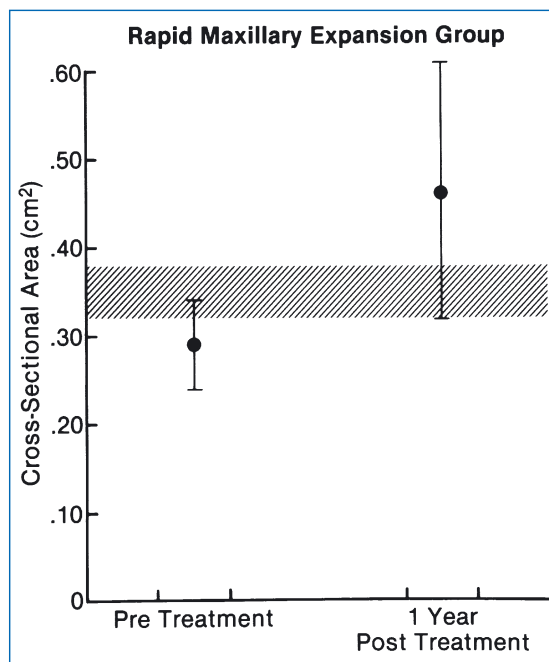
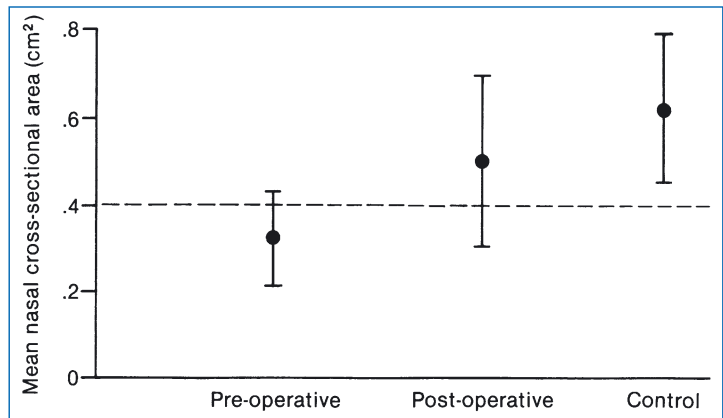


Fig. 33.7. Pre- and posttreatment data from the rapid maxillary expansion group

Fig. 33.8. Pre- and postoperative surgery data compared with normal adult group



ance nasal breathing, thereby modifying a long-term habit. Interestingly, this change in oral behavior does not occur when bone plates are used instead of prolonged maxillomandibular fixation.

33.6 Effects of the Nasal Airway on Speech

Speech is a modified breathing behavior that uses the respiratory system to provide an energy source and involves structures within the respiratory tract to modulate this energy into meaningful sounds. The oral, nasal, and pharyngeal structures that are affected by cleft lip and palate during breathing are often compromised for speech as well.

Cleft palate speech is usually characterized by two major distortions, one of resonance and the other of articulation [43–47]. Both relate to an inability to attain adequate velopharyngeal closure [48–51]. The articulation errors associated with velopharyngeal inadequacy are of particular interest to clinicians and researchers because the problems they pose usually remain after surgical repair, and the reasons they appear may have important implications for speech-motor control. It is paradoxical that, although velopharyngeal inadequacy stimulates compensatory speech behaviors, such responses tend to undermine, rather than enhance, speech performance.

Several compensatory strategies are used by cleft palate speakers [2, 52, 53]. Individuals with velopharyngeal inadequacy use greater respiratory effort or air volumes during speech. Their volumes are approximately twice those of normal speakers. The two factors responsible (i.e., airflow rate and duration of production) are both increased in cleft palate speakers. The implications of this are of interest because of the relationship between respiratory effort and speech performance. For example, in the presence of velopharyngeal

inadequacy, the use of larger volumes of air increases nasal emission. However, the relationship between nasal emission, voice quality, and sound intelligibility is complex and determined to an extent by the magnitude of nasal airway resistance. As discussed previously, nasal resistance is considerably higher in the cleft population [2]. This suggests that certain cleft palate speakers can compensate somewhat for velopharyngeal inadequacy by increasing respiratory efforts because high nasal resistance should raise intraoral pressure. On the other hand, nasal turbulence also occurs when airway resistance is high. Thus, an individual with nasal obstructions could conceivably produce undesirable turbulent noises and thereby decrease intelligibility with greater respiratory effort.

Compensatory changes in speech timing and alterations in tongue carriage also occur among cleft palate speakers [29, 54]. Apparently the level of intelligibility attained by cleft speakers is determined by the manner in which the various articulatory structures of the vocal tract respond to inadequacy, rather than the specific degree of impairment.

An important question among clinicians who treat speech problems associated with cleft palate is: why do compensatory speech behaviors that undermine performance develop? Recent studies suggest that these behaviors may be attempts to satisfy the requirements of a regulating system [28, 55–58]. The production of speech requires a relatively constant subglottal pressure which serves as the energy source [59–62]. Intraoral pressures for consonants also appear to be fairly constant within subjects although they do vary according to speaker age [63, 64] and sex [63, 65, 66]. In addition, they have been found to vary by word position [67–70], voicing characteristics [62, 71–77], vowel context [78–80], utterance length [70, 81, 82], and syllabic stress [82, 83]. Among cleft palate speakers, although peak intraoral pressure falls as the

extent of velopharyngeal impairment increases, it usually remains above 3 cm H₂O [3, 85]. Comparison of these findings with model simulations [84, 86] suggests that speakers make adjustments to velar impairment that tend to maintain pressure at levels thought to be necessary for obstruent consonant productions. A recent study [85] indicated that the ability to achieve adequate speech pressures depends on total upper airway resistance, that is, successful maintenance of consonant pressures depends on velar and nasal resistance. Specifically, individuals with velopharyngeal impairment utilize different respiratory volumes that are adjusted somewhat to the magnitude of upper airway resistance.

On the basis of these studies, Warren has suggested that speech aerodynamics follow the rules of a regulating system [58]. Specifically, for speech aerodynamics, subglottal pressure is kept relatively constant by checking or enhancing elastic forces and by compensating for sudden changes in respiratory load that occur with opening and closing of the upper airway. Precise control over the movement of upper airway structures and airflow tend to keep subglottal pressures regulated [57]. There is evidence that movements of the speech structures follow patterns that result in generally controlled vocal tract resistance and stable intraoral pressures [58].

The significance of a regulating system for cleft palate speakers relates to the loss of velar resistance with inadequate palate function. If nasal resistance is low, upper airway resistance could be supplemented by articulatory adjustments that increase airway resistance at other levels of the vocal tract. This implies that articulatory gestures may be modified by aerodynamic needs, and speech performance may be compromised. Indeed, a recent study by Peterson-Falzone [87] tends to support this assumption. Peterson-Falzone found that type of cleft affects the prevalence of compensatory articulation errors associated with velopharyngeal impairment. Errors were more frequent for subjects with bilateral cleft lip and palate, followed by cleft palate only, and least frequent in individuals with unilateral cleft lip and palate. Interestingly, earlier in this chapter we reported that children with unilateral cleft lip and palate had the most compromised airways and those with bilateral clefts of the lip and palate were the least compromised. This supports the thesis that articulatory patterns may be modified by aerodynamic needs. The pharyngeal fricative, glottal stop, nasal grimace, and high tongue carriage may be examples of such responses. Speakers increase airway resistance, but undermine intelligibility. Whether these behaviors are prompted by low nasal resistance is a question being actively pursued at the present time.

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34 Surgical Management of Velopharyngeal Dysfunction

RICHARD E. KIRSCHNER, RACHEL A. RUOTOLO

The production of normal speech is dependent upon the structural and functional integrity of the velopharyngeal valve. Inability to uncouple the oral and nasal cavities during speech is manifest as hypernasal resonance and nasal air emission and may result in the development of compensatory articulation errors which further compromise speech intelligibility. Velopharyngeal dysfunction (VPD) is most often caused by overt or submucosal clefts of the secondary palate but may also be diagnosed in patients with congenital or acquired neuromuscular disorders and in those who have undergone adenoidectomy. Other patients may manifest congenital abnormalities of velopharyngeal function either associated or unassociated with known genetic disorders.

The goal of surgical intervention is to produce a competent velopharyngeal mechanism for speech while avoiding the induction of nasal airway obstruction. Ideally, postoperative speech is characterized by the presence of normal resonance and the absence of audible nasal air escape. Elimination of hypernasality is itself, however, an insufficient measure of surgical success. The postoperative appearance of nasal airway obstruction, as evidenced by hyponasal speech, obligate mouth breathing, and/or obstructive sleep apnea, should be considered an unacceptable outcome.

Surgical management of VPD thus represents a balance between persistent hypernasality on one hand and nasal airway obstruction on the other. Successful management, therefore, depends upon selecting the most appropriate surgical procedure for each patient based upon a careful analysis of the patient's history and velopharyngeal anatomy and dynamics. All surgical procedures for the treatment of VPD seek to improve velopharyngeal valving by reducing the cross-sectional area of the velopharyngeal port. The procedures employed most commonly today are the posterior pharyngeal flap, the sphincter pharyngoplasty, and the Furlow double-opposing z-palatoplas-

ty. Posterior pharyngeal wall augmentation is a technique employed less frequently.

34.1 Patient Evaluation

All patients with suspected VPD should undergo a thorough history, a comprehensive speech and language evaluation, and a careful physical examination. It is essential to know the details of all prior surgical procedures, such as palatoplasty, prior pharyngoplasty, and tonsillectomy and/or adenoidectomy, that may influence velopharyngeal function. If the levator muscles remain sagittally oriented, consideration should be given to performing a palatal pushback procedure with intra-velar veloplasty or a Furlow palatoplasty either alone (small gap VPD) or in combination with a pharyngoplasty (large gap VPD). Persistent VPD despite prior pharyngoplasty warrants particular care in diagnosis and surgical management in order to eliminate ongoing hypernasality while avoiding the complication of upper airway obstruction. Previous tonsillectomy should be noted, as such may result in scarring of the posterior tonsillar pillars, rendering sphincter pharyngoplasty problematic in some patients.

Surgical management may be complicated by the presence of comorbid conditions, particularly in syndromic patients (e.g., cardiac anomalies in patients with 22q11.2 deletion syndrome). Appropriate medical and anesthetic consultation should be obtained preoperatively in all such patients. Those with a history of airway compromise, such as patients with Pierre Robin sequence, or who present with loud snoring or other evidence of upper airway obstruction require a thorough preoperative investigation of the upper airway as well as a polysomnogram to rule out the presence of obstructive sleep apnea.

The diagnosis of VPD is confirmed in all patients by nasendoscopy and/or multiview videofluoroscopy

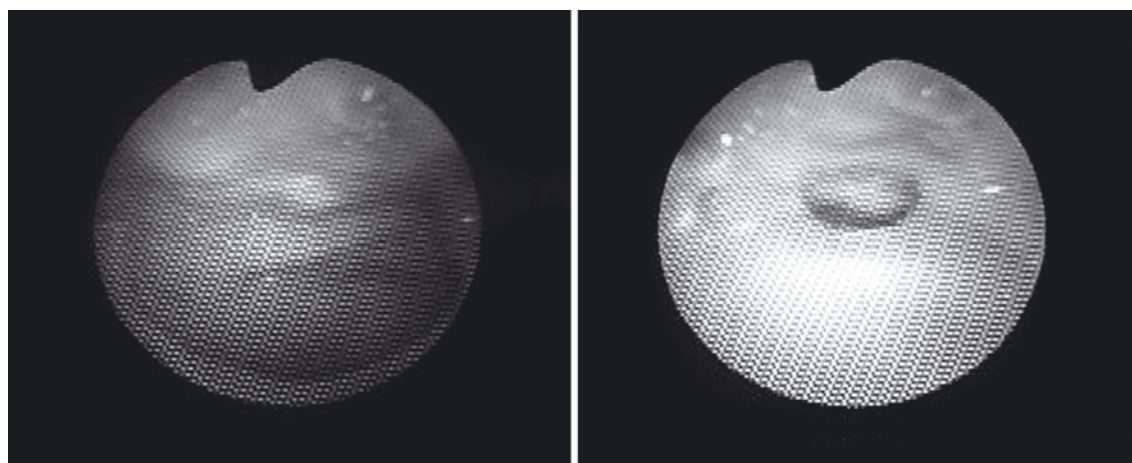


Fig. 34.1. Nasoendoscopic images of the velopharyngeal structures in a child with VPD. *Left*, Velopharyngeal opening at rest. *Right*, Persistent velopharyngeal gap during production of “sss” sound

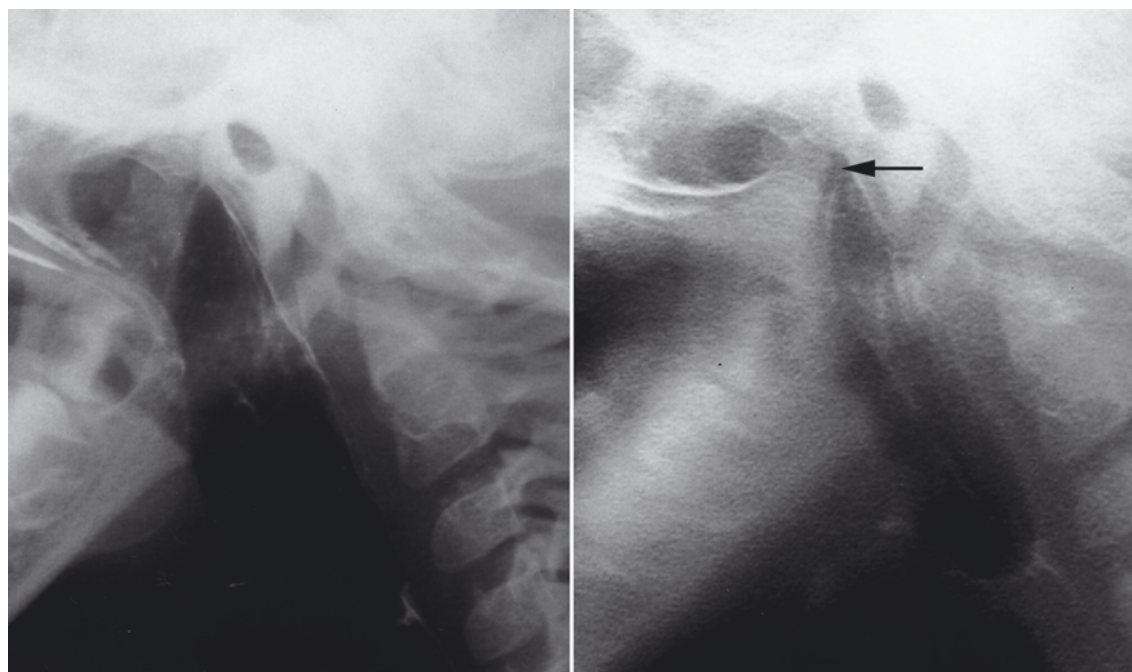


Fig. 34.2. Videofluoroscopic images of the velopharyngeal structures in a child with VPD. *Left*, Velopharyngeal opening at rest. *Right*, Persistent velopharyngeal gap (arrow) during production of “puppy.” (From [84] with permission)

(Figs. 34.1, 34.2). Although either study alone will provide ample information to establish the diagnosis, the two studies provide complimentary data with respect to velopharyngeal anatomy and function. The site, pattern, and symmetry of velopharyngeal closure should be noted, as should gap size and location, velar muscular anatomy, tonsillar size, and adenoid mor-

phology. Hypoplastic adenoids may contribute to VPD in patients with 22q11.2 deletion syndrome [16, 25, 75], and affected patients are best managed by creation of a posterior pharyngeal flap or sphincter pharyngoplasty. On the other hand, enlarged tonsils or irregularly shaped adenoids may impair velopharyngeal valving in some nonsyndromic patients [42,

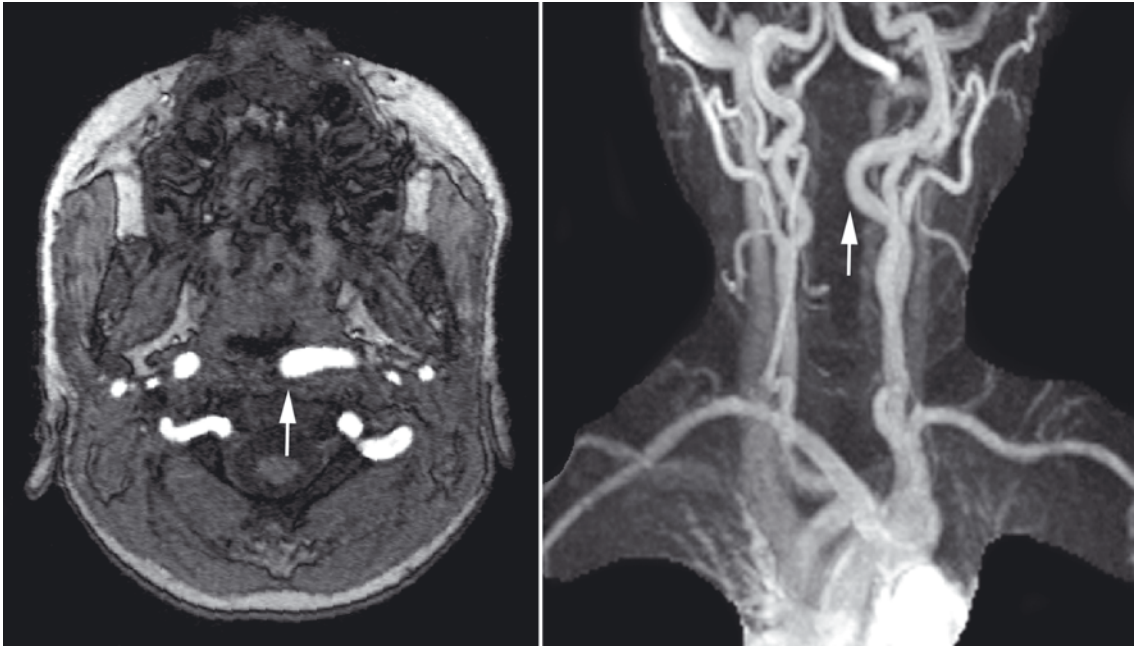


Fig. 34.3. Medial deviation of the internal carotid artery (arrow) at the level of the nasopharynx in a child with a chromosome 22q11.2 deletion. (From [84] with permission)

52, 61]. Preoperative imaging studies to determine the contribution of the tonsils and adenoids to velopharyngeal function are essential in such patients in order to avoid misguided surgery. For these patients, tonsillectomy and/or adenoidectomy is the proper initial management

Magnetic resonance imaging should be performed preoperatively in all patients with 22q11.2 deletion syndrome, since one in five such patients will manifest medial displacement of one or both internal carotid arteries [12, 43, 44, 55] (Fig. 34.3). In some cases, the displaced carotid artery may approach the midline in the retropharyngeal space at the level of the first cervical vertebra, placing the vessel at risk for injury during the course of pharyngoplasty. Arterial pulsations noted at the time of endoscopy are frequently transmitted through the surrounding soft tissues, rendering endoscopy alone an unreliable means of assessing carotid anatomy. Although some have suggested that carotid artery displacement should be considered a contraindication to pharyngoplasty, others have demonstrated that surgery may indeed be safely performed in such patients [66, 78]. In many cases, the displaced vessel will lateralize with neck extension at the time of surgery. When the vessel can still be palpated deep the posterior pharyngeal wall after positioning, site of a posterior pharyngeal flap can be modified to avoid exposure of the vessel. In all such cases, the use of preoperative carotid imaging

plays an important role in surgical planning and informed consent.

34.2 Posterior Pharyngeal Flap

The posterior pharyngeal flap functions as a soft tissue obturator of the velopharyngeal port. Closure of the lateral openings beside the sagittally oriented flap is primarily achieved by medial movement of the lateral pharyngeal walls [4]. Thus, use of a pharyngeal flap is best suited to those patients who demonstrate good lateral pharyngeal wall motion, as in a sagittal or circular velopharyngeal closure pattern. Passavant first reported secondary management of VPD by simple adhesion of the velum to the posterior pharynx in 1865 [47]. Elevation of an inferiorly based flap from the posterior pharyngeal wall was described by Schoenborn in 1875 [57]. In 1886, he reported on the use of a superiorly based flap [58.]. The superiorly based flap was described in the USA by Padgett in 1930 [46]. In 1973, Hogan [29] advocated the intraoperative use of rubber catheters to allow for creation of a wide pharyngeal flap with “lateral port control.” In his technique, port size was based upon the pressure-flow studies of Warren et al. [71, 72], wherein velopharyngeal valve area of greater than 20 mm² was documented to result in hypernasality and nasal air escape during connected speech. Although Hogan reported

excellent results in his series, many have since abandoned the use of “lateral port control,” citing a greater potential for postoperative airway obstruction.

The selection of either an inferiorly or a superiorly based flap remains the subject of some debate. While technically easier to perform, the inferiorly based flap has the disadvantages of limited flap length and an obscured donor site. The inferiorly based flap also has the theoretical disadvantage of tethering of the soft palate inferiorly, although a poorly designed superiorly based flap may also result in such tethering. Several studies have failed to document a significant difference in surgical outcome between patients who have had superiorly based flaps and those who have had flaps based inferiorly [24, 63, 74]. Nevertheless, the superiorly based flap remains more commonly used today.

34.2.1 Surgical Technique (Fig. 34.4)

The superiorly based pharyngeal flap, composed of pharyngeal mucosa and the underlying superior pharyngeal constrictor muscle, is elevated from the pre-

vertebral fascia and inset into the posterior velum. In order to optimize surgical outcome, flap design should be individualized, based upon anatomic and functional information provided by preoperative imaging studies. Ideally, the base of flap should correspond to the level of velopharyngeal closure, high on the posterior pharyngeal wall. When there is a marked asymmetry in lateral pharyngeal wall motion, or when there is significant displacement of a carotid artery, the origin and inset of the flap may be skewed to one side [3]. Flap width is determined by the size of the gap to be obturated. Care should be taken to avoid creation of side ports that are too small, however, as such may lead to nasal airway obstruction and to sleep apnea.

Inset of the pharyngeal flap into the nasal surface of the velum may be accomplished either by dividing the soft palate in the midline or by dissecting a sub-mucosal pocket through a posterior transverse, or “fishmouth,” incision. Use of the latter allows for fairly precise control of flap width, as described by Argamaso [2]. Use of a broad, relatively short flap avoids the tendency of unlined flaps to “tube” upon themselves and to thereby produce a narrow flap with large

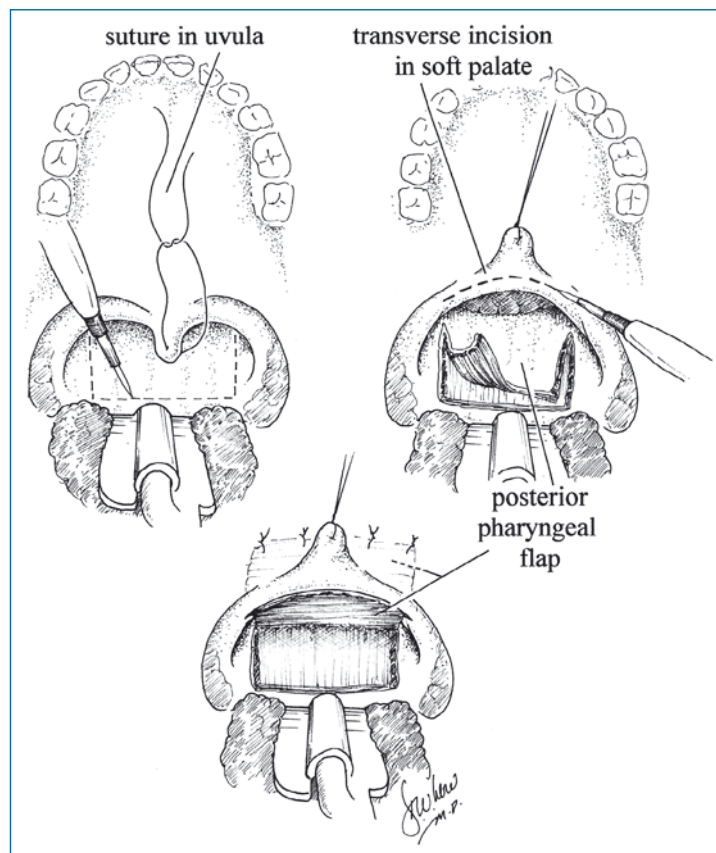


Fig. 34.4. Posterior pharyngeal flap. (From [84] with permission)

side ports [69]. The raw oral surface of narrower flaps should be lined by turnover flaps of soft palatal mucosa to avoid this otherwise inevitable outcome. The donor site of narrow flaps and those of moderate width may be closed primarily. When use of a wide flap precludes donor site closure, the posterior pharyngeal wall may be left to heal secondarily within 2–3 weeks.

34.2.2 Outcome

Decades of successful use have established the posterior pharyngeal flap as the “gold standard” for surgical management of VPD. Accurate interpretation of reported series is hindered by the use of different surgical techniques, heterogeneous patient characteristics, and nonstandardized measures of surgical outcome. Argamaso [2] reported elimination of hypernasality in 96.1% of 226 patients after pharyngeal flap surgery. In a recent long-term outcomes study of pharyngeal flap surgery at the University of Iowa [9], investigators rated hypernasality on a scale from 1 to 6, with a score of 1 indicating no hypernasality and that of 6 denoting severe hypernasality. Mean scores were 1.94, 1.53, 1.71, and 1.75 after 2- to 5-year, 5- to 8-year, 8- to 11-year, and 11- to 14-year follow-up, respectively, indicating that pharyngeal flap surgery most often significantly improves or eliminates hypernasality and that the surgical outcome is durable.

Complications of pharyngeal flap surgery include bleeding, dehiscence, nasal airway obstruction, and obstructive sleep apnea [28]. Rare cases of postoperative death have been reported and are primarily the result of upper airway obstruction [68]. Fraulin et al. [17] reported that the surgeon involved, the presence of associated medical conditions, concurrent surgical procedures, and an open donor site may be predictive factors for complications. Bleeding most often occurs within the first 24 h and may be reduced by closure of the donor site and by the use of electrocautery to elevate the flap. Although many patients will demonstrate some degree of upper airway obstruction in the early postoperative period, this most often resolves as edema subsides. Syndromic patients and those with a history of Pierre Robin sequence may present with structural narrowing of the upper airway or pharyngeal hypotonia and may therefore be at greater risk for postoperative obstructive sleep apnea [1, 73]. Wells et al. [73] reviewed their 17-year experience with 111 patients who underwent posterior pharyngeal flap surgery for management of VPD. Twelve patients demonstrated clinical evidence of postoperative nocturnal respiratory obstruction, and three required takedown of the flap. Nine of 12 patients with obstructive symptoms underwent polysomnographic evaluation, eight

of which were normal. Thus, although nocturnal respiratory obstruction may occur commonly after pharyngeal flap surgery, obstructive symptoms do not necessarily imply the occurrence of obstructive sleep apnea. In order to determine the incidence of obstructive sleep apnea after pharyngeal flap surgery, Sirois et al. [62] evaluated postoperative polysomnograms in 40 patients. Abnormal studies were obtained in 14 patients (35%), of whom six demonstrated obstructive apnea, six central apnea, and two mixed obstructive and central apnea. Ten of the 14 patients were restudied in the following months, eight of whom demonstrated normal studies and two of whom demonstrated central apnea. The remaining four patients were noted to demonstrate no obstructive symptoms. Patients with tonsillar hypertrophy may be at greater risk for postoperative obstructive apnea and should therefore undergo tonsillectomy either prior to or at the time of pharyngeal flap surgery [82].

Postoperative monitoring of the upper airway, including continuous oximetry, plays an important role in the management of all patients undergoing pharyngeal flap surgery. Use of a nasopharyngeal airway and admission to an intensive care unit should be considered for those patients at highest risk for postoperative airway obstruction. Patients are discharged once they demonstrate adequate airway stability and oral intake of fluids.

34.3 Sphincter Pharyngoplasty (Fig. 34.5)

In 1950, Hynes described a technique for managing VPD after palatoplasty by horizontal transposition of myomucosal flaps containing the salpingopharyngeus muscles [33]. He later described inclusion of the palatopharyngeus muscles in the flaps and stressed the need to make the flaps as bulky as possible in order to optimize speech outcome [34, 35]. As originally described, the Hynes pharyngoplasty was a two-stage procedure designed to narrow the velopharynx. Orticochea [45] first described the construction of a “dynamic sphincter” by inset of bilateral palatopharyngeal myomucosal flaps into an inferiorly based flap on the posterior pharynx at a level somewhat below that of attempted velopharyngeal closure. Jackson and Silverton [36] later modified the technique by use of a superiorly based flap for inset of the flaps.

Riski et al. [54] have stressed the need to inset the flaps high on the posterior pharyngeal wall at the level of attempted velopharyngeal closure as determined by preoperative imaging studies. Since a dynamic sphincter may be achieved in only two thirds of patients [37], anatomic factors, such as reduced port size and augmentation of the posterior pharyngeal wall, may be of critical importance in determining outcome.

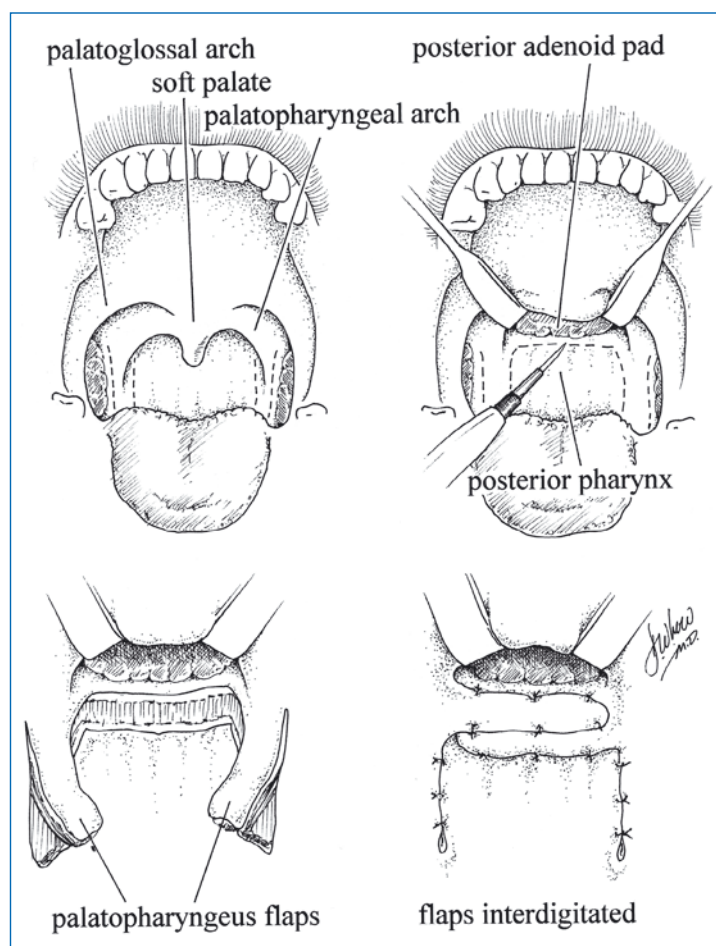


Fig. 34.5. Sphincter pharyngoplasty. (From [84] with permission)

34.3.1 Surgical Technique

The surgical technique used most widely today represents a modification of the Hynes pharyngoplasty. Bilateral superiorly based palatopharyngeal flaps are inset into a simple transverse incision in the posterior pharyngeal wall at the level of velopharyngeal closure. The level of flap inset is limited superiorly by the adenoid pad, since the friable adenoidal mucosa bleeds easily and provides little strength for holding sutures. Once elevated, the palatopharyngeal flaps are turned 90°, crossing one another, and their tips are sutured into the contralateral corners. Suture of the flaps to the posterior pharynx and to one another creates a transverse mound of tissue on the posterior pharyngeal wall and a small central velopharyngeal port.

34.3.2 Outcome

In a retrospective review of speech outcome after sphincter pharyngoplasty, Shewmake et al. [60] reported that 85.4% of 48 patients demonstrated normal resonance, 8.3% demonstrated mild residual VPD, 4.2% demonstrated moderate residual VPD requiring secondary tightening of the port, and 2.1% demonstrated hyponasality. Riski et al. [53] reported resolution of hypernasality and normal pressure flow measurements in 78% of 139 patients, the failures noted to be primarily the result of the flaps being inset below the level of attempted velopharyngeal closure. In a review of 123 patients, Witt et al. [79] noted that 16% of sphincter pharyngoplasties required revision, the primary etiology of failure being partial or complete flap dehiscence. Losken et al. [41] reviewed

their experience with 250 patients, noting a 12.8% revision rate, noting that failure to achieve normal resonance was most common in patients with VPD associated with a chromosome 22q11.2 deletion. Patients who required revision demonstrated significantly greater preoperative measures of nasalance and velopharyngeal valve area. Despite relatively consistent reported speech outcomes after sphincter pharyngoplasty, however, it remains difficult to predict which patients are at greatest risk for failure.

The question of whether or not a dynamic sphincter is created by sphincter pharyngoplasty procedures remains the subject of some debate. Witt et al. [76] examined pre- and postoperative videofluoroscopic examinations in 20 patients undergoing sphincter pharyngoplasty, noting that all patients exhibited some dynamism, although there was a wide variation noted in the extent of muscular activity of the sphincter. In contrast, Kawamoto [37] reported that while postoperative nasendoscopy demonstrated complete velopharyngeal closure in 89% of 18 patients undergoing sphincter pharyngoplasty, a dynamic sphincter was achieved in only 67%. Thus, anatomical factors, such as reduction in port size and augmentation of the posterior pharyngeal wall, may play an important role in surgical outcome.

Complications of sphincter pharyngoplasty include flap dehiscence, nasal airway obstruction, and obstructive sleep apnea. Sphincter pharyngoplasty is considered by its advocates to be more “physiologic” than the posterior pharyngeal flap and therefore less likely to induce postoperative airway dysfunction. There is substantial evidence however, that sphincter pharyngoplasty may also result in nasopharyngeal airway obstruction, as manifested by hyponasality and disrupted sleep architecture. Witt et al. [77] reported postoperative airway dysfunction in eight of 58 patients who underwent sphincter pharyngoplasty, five of whom had been previously diagnosed with Pierre Robin sequence. Obstructive signs resolved in all but two patients within 3 days of surgery, and the remaining patients were successfully managed with continuous positive airway pressure (CPAP). No patient required takedown of the sphincter. Saint Raymond et al. [56] examined polysomnographic studies in 17 patients before and after modified Orticochea pharyngoplasty. They found that the procedure induced no significant impairment in either the apnea-hypopnea index or the nocturnal oxygen saturation. Nevertheless, there was a reduction in slow wave sleep and an increase in the number of cortical microarousals postoperatively, suggesting that reduction in nasopharyngeal airway diameter after sphincter pharyngoplasty may increase respiratory effort sufficiently enough to cause sleep fragmentation even in the absence of obstructive apnea.

34.3.3 Comparison of Posterior Pharyngeal Flap and Sphincter Pharyngoplasty

There have been few studies which compare the results of posterior pharyngeal flap procedures with those of sphincter pharyngoplasty. Pensler and Reich [49] retrospectively compared 75 patients with VPD treated by pharyngeal flap to 10 treated by sphincter pharyngoplasty, noting no significant difference in speech outcome. Three patients in the series demonstrated postoperative sleep apnea, all of whom had been treated by pharyngeal flaps. Witt et al. [79] examined velopharyngeal function in 65 patients after pharyngeal flap surgery and in 123 patients after sphincter pharyngoplasty. Persistent VPD requiring surgical revision was present in 20% of pharyngeal flap patients and 16% of sphincter pharyngoplasty patients. In the only prospective randomized series reported to date, Ysunza et al. [83] treated 50 patients with VPD after palatoplasty with either pharyngeal flap procedures or sphincter pharyngoplasty. The frequency of residual VPD after velopharyngeal surgery was not significantly different between the groups (12% vs. 16%, respectively). The authors stressed, however, the importance of customizing both procedures to the findings of preoperative imaging studies of each patient. Moreover, there appears to be an inverse relationship in most patients between postoperative symptoms of upper airway obstruction and residual VPD, suggesting that velopharyngeal competence may occasionally come at the cost of airway obstruction, usually transient, particularly in patients undergoing pharyngeal flap procedures.

34.4 Furlow Double Opposing Z-Palatoplasty (Fig. 34.6)

Although originally designed as a technique for the repair of palatal clefts, the double-opposing z-plasty has received considerable attention as a treatment option for VPD. The technique, first described by Leonard Furlow in 1978 [19], offers several theoretical advantages for the management of VPD in selected patients. Transposition of the posteriorly based myomucosal flaps reorients the levator veli palatini muscles, reconstructing the levator sling. The z-plasty lengthens the velum while reducing the potential for velar shortening otherwise associated with a midline longitudinal scar. Thus, the repair is suitable for patients with small- to moderate-gap velopharyngeal insufficiency associated with sagittal orientation of the levators, such as those with a submucosal cleft palate or those who have undergone prior palatoplasty without veloplasty. The procedure should not be performed in patients with an anatomically normal

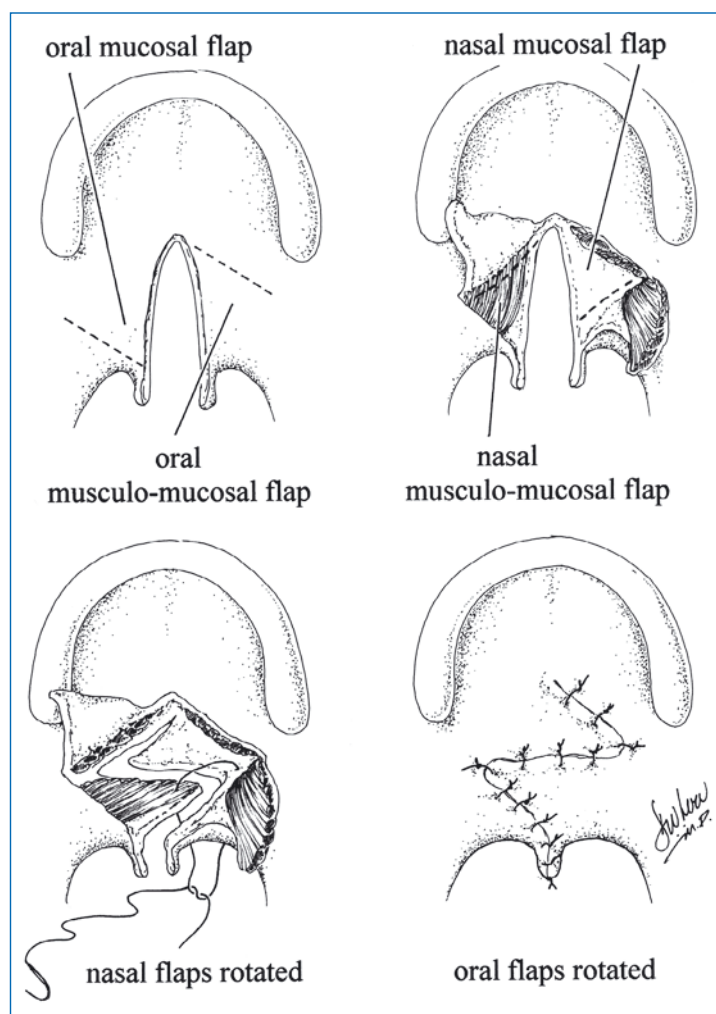


Fig. 34.6. Furlow double opposing z-palatoplasty. (From [84] with permission)

palate or in those who have undergone prior surgical reconstruction of the levator sling.

34.4.1 Surgical Technique

The Furlow palatoplasty comprises mirror image z-plasties of the oral and nasal velar mucosa. The posteriorly based triangular flaps are dissected as myomucosal flaps; the anterior flaps are of mucosa and submucosa alone.

The oral z-plasty incisions extend from the hamulus to the junction of the hard and soft palate in the midline on one side and from the base of the uvula in the midline to the hamulus on the contralateral side. The central limb of the z-plasty joins the lateral limbs along the midline of the velum. The posteriorly based oral myomucosal flap is elevated in the nasal submucosal plane, separating the velar muscles from the

midline and from the posterior edge of the hard palate. Dissection should proceed laterally to the hamulus, and the palatal aponeurosis must be completely divided in order to allow for complete rotation of the flap and repositioning of the levator muscle. Incomplete division of the palatal aponeurosis results in anterior tethering of the levator. Once the oral myomucosal flap has been elevated the anteriorly based nasal mucosal flap may be incised from the base of the uvula to a point just medial to the eustachian tube orifice.

On the contralateral side, the anteriorly based mucosal flap is elevated in the oral submucosal plane. The posteriorly based nasal myomucosal flap is then divided along the posterior edge of the hard palate. Again, care must be taken to ensure complete division of the palatal aponeurosis in order to allow for posterior rotation of the flap and for complete reorientation and repositioning of the levator muscle. The

nasal flaps are then transposed and sutured, followed by the oral flaps. Transposition of the posteriorly based flaps overlaps the levator muscles and reconstructs the levator sling.

34.4.2 Outcome

Huang et al. [31] compared cephalometric radiographs from patients who had undergone Furlow palatoplasty to those of age- and gender-matched controls. Mean velar length in the Furlow group was greater than that of cleft patients who underwent non-Furlow repair, but not significantly different from that of noncleft controls. These results suggest that palatal lengthening does indeed occur in some patients following double opposing z-plasty and that such may result in normal or near-normal velar length. The authors also documented greater velar thickness in some patients after Furlow palatoplasty, a finding likely attributable to overlap of the velar muscles. Others have also verified that palate repair by double-opposing z-plasty results in velar lengthening. D'Antonio et al. [13] prospectively studied changes in radiographic dimensions and aerodynamic measures in twelve patients with repaired cleft palate or unrepaired submucosal cleft palate, all of whom demonstrated VPD. Although mean velar length and mean velar thickness were greater postoperatively, not all patients demonstrated significant alterations in palatal dimensions, and not all patients achieved velopharyngeal competence. Patients who achieved velopharyngeal closure after surgery, however, demonstrated greater increases in velar length, velar thickness, or both than those who demonstrated persistent abnormalities in velopharyngeal valving. Thus, speech outcome after Furlow double-opposing z-plasty may be determined in large part by the extent of palatal lengthening achieved. This, in turn, is determined in part by the angles of the z-plasty. It is important to note, however, that these angles are based not upon any specific geometric design, but are determined by the anatomy of the palate in each patient. As Randall et al. have noted, the z-plasty angles may approach 90° in patients with a short velum [50]. Less than optimal results after Furlow z-plasty may therefore be related to the constraints imposed by palatal anatomy. On the other hand, strict adherence to prescribed flap angles may result in technical errors in flap dissection which themselves may compromise surgical outcome [20].

Available data suggest that Furlow double-opposing z-plasty may be an effective technique for the management of VPD in selected patients after primary cleft palate repair or in patients with submucosal cleft palate. Chen et al. [11] reported that 16 of 18 patients with VPD after palate repair achieved

velopharyngeal competence after velar revision by Furlow z-plasty. All but one of these patients had a velopharyngeal gap of less than 5 mm preoperatively. In contrast, the two patients with persistent VPD had gaps of greater than 10 mm. Similarly, Hudson et al. [32] noted that 85% of patients with VPD after primary palatoplasty demonstrated normal resonance after Furlow repair. D'Antonio et al. [13] reported that Furlow z-plasty resulted in velopharyngeal competence in six of eight patients with persistent VPD after cleft palate repair. All patients in the series demonstrated endoscopic findings of a central V-shaped notch in the posterior velum, good velar motion, and a "small" velopharyngeal gap.

Seagle et al. [59] reported a successful outcome after Furlow repair in 83% of patients with VPD associated with a submucosal cleft palate, noting a high likelihood of success when the velopharyngeal gap measured less than 8 mm preoperatively and poorer results with gaps larger than 8 mm. Chen et al. [10] demonstrated a 96.7% velopharyngeal competency rate after submucosal cleft repair in patients with a gap of less than 5 mm. In contrast, D'Antonio et al. [13] reported that velopharyngeal competence was achieved in just one of four patients who underwent Furlow z-plasty for repair of submucosal clefts. Although the reasons for this discrepancy are unclear, the authors note that the three patients with persistent VPD demonstrated only small increases in velar length and thickness postoperatively.

Complications of Furlow double-opposing z-plasty include bleeding, oronasal fistula, and nasopharyngeal airway obstruction. Liao et al. [39] compared postoperative airway compromise in 20 patients who underwent Furlow palatoplasty for management of VPD with that in 28 patients who underwent pharyngeal flap surgery. Polysomnograms performed 6 or more months after surgery documented a significantly lower incidence and severity of obstructive sleep apnea in the Furlow group. In an earlier study, the same authors found polysomnographic evidence of mild obstructive apnea in all patients who underwent Furlow repair, although this resolved in nearly all patients within 3 months [40].

34.5 Posterior Pharyngeal Wall Augmentation

Surgeons have long sought to improve velopharyngeal closure in selected patients with VPD by augmenting the posterior pharyngeal wall in order to diminish the sagittal dimension of the velopharyngeal orifice. The literature is replete with references to the use of both implantable and injectable materials, both autologous/homologous and alloplastic. Reported results

have been variable, depending upon the patient population studied and the implant material used. In general, the best results have been observed in patients with good velar motion and relatively small velopharyngeal gaps.

In 1879, Passavant described using a single-pedicle soft tissue flap folded over itself and inset on the posterior pharyngeal wall [48]. This soft tissue augmentation proved to be transient, however, and the technique was quickly abandoned. In 1900, Gersuny reported augmentation of the posterior pharyngeal wall by injection of petroleum jelly [22]. Although that technique improved the speech of many patients, serious complications, including blindness and death, were later reported. Eckstein later reported the use of paraffin injections without the untoward complications seen with petroleum jelly [15]. In 1912, Hollweg and Perthes proposed the use of autologous cartilage grafts placed through a transcervical approach to augment the posterior pharynx [30]. They reported positive outcomes, leading others to attempt variations on their published technique. Wardill [70], Lando [38], and Hess [26] reported significant speech improvement in patients treated with cartilage grafts placed through a transoral approach. Bently [6], however, reported inadequate speech improvement 8 years following this procedure. In order to avoid the complication of graft resorption following the use of autologous and homologous cartilage, Blocksma investigated the use of Silastic implants for pharyngeal wall augmentation [7]. He hypothesized that silicone augmentation pharyngoplasty would prove to be a simpler, less expensive procedure that produced more reliable, longer-lasting results. His experience revealed a high incidence of implant infection and extrusion, however, leading him to conclude that autologous implants should be considered the preferred augmentation material for the treatment of VPD.

In 1968, Bluestone et al. reported success after injection of Teflon into the posterior pharynx, noting no instances of infection, extrusion, or foreign body reaction [8]. They emphasized that this technique was suitable primarily for those patients with good levator function and small velopharyngeal gaps as determined by preoperative imaging. These findings were later confirmed by Sturim and Jacob [65], the only negative sequela reported being that of neck stiffness in a small number of patients. Other materials that have been used for pharyngeal wall augmentation with variable success include silicone, Proplast I [81], allograft cartilage [67], injectable collagen [51], and autologous fat and dermis.

34.5.1 Surgical Technique

Patient selection for posterior pharyngeal wall augmentation requires careful examination of gap size, velar mobility, and velopharyngeal closure pattern. The material to be used for augmentation is selected, and the exact site for graft placement is determined by preoperative imaging studies. The anterior tubercle of the atlas is identified and palpated. A local anesthetic agent containing epinephrine is infiltrated into the posterior pharynx, facilitating a hemostatic dissection. A transverse or vertical incision is then made approximately 2 cm below the site selected for implant placement. Dissection with scissors or electrocautery creates a small pocket for the graft between the superior constrictor muscle and the prevertebral fascia. Care must be taken to place the graft material in the precise position of attempted velopharyngeal closure, usually just above the tubercle of the atlas, and to carefully secure the graft to prevent its inferior displacement.

If the material chosen for augmentation is injectable, the same landmarks are identified, and the material is injected at the level of attempted velopharyngeal closure. The material should be placed in the submucosal plane and care should be taken to avoid injection into the prevertebral fascia [21].

34.5.2 Outcome

Smith and McCabe reported resolution of hypernasality in 60% and moderate improvement in 18% of 80 patients who underwent Teflon augmentation pharyngoplasty for management of VPD [64]. All patients demonstrated good preoperative palatal motion. Furlow et al. reported resolution of hypernasal speech in 74% of 35 patients who underwent Teflon injection for posterior pharyngeal wall augmentation, including eight patients with failed pharyngeal flaps [21]. Despite these favorable results, however, efforts to obtain FDA approval for the use of Teflon for the management of VPD have ceased.

Wolford et al. reported the use of Proplast I in 26 patients [81]. Although they reported implant loss in four patients and dislodgment in two patients, they advocated its use in young patients and those with small VP gaps. Injectable collagen was introduced for posterior pharyngeal wall augmentation by Remacle et al., who reported favorable results with no complications in a limited study of only five patients [51].

Denny et al. reported that velopharyngeal competence was achieved in 25% of 20 patients with VPD treated by retropharyngeal implantation of costal bone or cartilage grafts and improvement in speech without elimination of VPD in another 65% [14]. Follow-up studies of the same cohort, however, failed to support the durability of these results [80]. In 1997, Witt et al. reported on autologous posterior pharyngeal wall augmentation using a rolled superiorly based pharyngeal myomucosal flap, noting no detectable improvement in VPD in the 14 patients studied [80]. Gray et al., however, reported that a folded flap to augment the posterior wall was just as likely to be effective as other surgical techniques to treat small velopharyngeal gaps [23]. Their outcomes were best in patients who were young and who had good velar motion.

In general, it is accepted that augmentation of the posterior pharyngeal wall is indicated in patients who have adequate mobility of their soft palate and who have small gaps as determined by preoperative radiologic and/or endoscopic studies. The procedure may have a role in patients with “borderline” VPD, in those with VPD after adenoidectomy, and in those considered to be at high risk for obstructive sleep apnea after pharyngeal flap or sphincter pharyngoplasty. Because no single alloplastic material has been found to be uniformly safe, effective, and reliable, and because of the variable reported success associated with the use of autologous tissue, this technique should be considered only as a secondary option for treatment of carefully selected patients with VPD.

34.6 Management of Persistent VPD

Although surgical management of VPD successfully eliminates hypernasality in most patients, approximately 15%–20% of posterior pharyngeal flap procedures and sphincter pharyngoplasty procedures will require revision for persistent VPD [79]. Persistent hypernasality after posterior pharyngeal flap surgery may be the result of flap dehiscence or may occur when lateral pharyngeal wall motion is insufficient to occlude the side ports during speech. The latter may result from poor flap design (too narrow), cicatricial flap narrowing, or poor patient selection (poor lateral wall motion) (Fig. 34.7). In some cases, a scarred flap that is located too low on the posterior pharyngeal wall may tether the velum, resulting in persistent VPD. Sphincter pharyngoplasty may fail as the result of dehiscence, poor design (too low) or incomplete closure of the central port.

Patients with perceptual evidence of persistent VPD after surgical treatment should undergo diagnostic imaging of the velopharyngeal port by

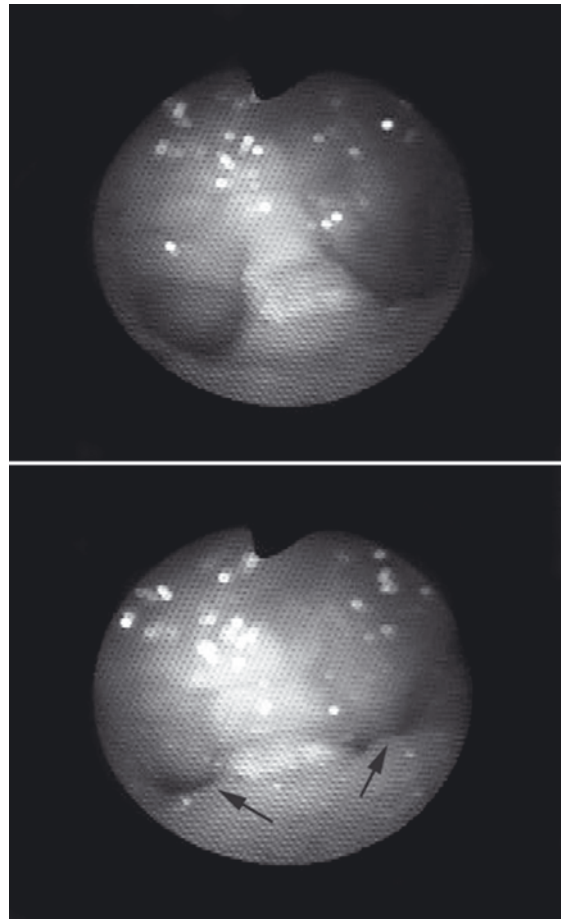


Fig. 34.7. Persistent VPD after posterior pharyngeal flap surgery. *Top*, Narrow pharyngeal flap at rest. *Bottom*, Persistent velopharyngeal gap (arrows) during phonation

nasendoscopy or multiview videofluoroscopy. All patients being considered for surgical revision should be carefully screened for evidence of upper airway obstruction, and revision should be reserved for those with a stable airway. The management of persistent VPD should be tailored to the patient's overall medical condition, airway status, and the findings of preoperative imaging studies. Posterior pharyngeal flaps that are found to be too narrow may be augmented by “patch” flaps, replaced by wider flaps elevated from the scarred posterior pharyngeal wall, or divided and replaced by a sphincter pharyngoplasty [5, 18, 27, 79]. Low flaps that tether the velum may be repositioned higher on the posterior pharyngeal wall by a V-Y advancement. Incomplete port closure after sphincter pharyngoplasty may be managed by port tightening or by the addition of a carefully tailored posterior pharyngeal flap. Although elevation of a pharyngeal

flap from a scarred posterior pharynx – especially after sphincter pharyngoplasty – has the theoretical disadvantage of poor blood supply to the flap, the procedure can indeed be performed safely 6–12 months after the primary operation [5].

Just as for primary surgical management, successful secondary treatment of VPD requires careful preoperative evaluation and individualized management. Barone et al. [5] reported on 18 patients who demonstrated bilateral port insufficiency after posterior pharyngeal flap surgery that was managed by elevation of a new superiorly based pharyngeal flap from the scarred posterior pharynx and three patients who demonstrated unilateral port insufficiency that was managed by inset of a “patch” flap. Postoperatively, 18 patients demonstrated normal resonance, two remained hypernasal, and one was hypernasal. Witt et al. [79] subsequently reviewed the results of salvage surgery for failed pharyngeal flaps in 13 patients, noting that eight were successfully managed with a single revisional procedure. The remaining five patients achieved velopharyngeal competence after a second revision, but all in this group developed hyponasal speech. In the same study, 17 of 20 failed sphincter pharyngoplasties were successfully salvaged after surgical revision.

34.6 Summary

There are many causes of impaired velopharyngeal valving that may occur in combination both in the presence and in the absence of overt or submucosal clefts of the palate. Moreover, there is a wide spectrum of severity of velopharyngeal dysfunction, and it is important to understand that such may be but one of several communication disorders that may contribute to impaired speech intelligibility in any given patient. Optimization of surgical outcome in each patient, therefore, requires both precision in diagnosis and careful selection of surgical technique.

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35 Velopharyngeal Dysfunction Management Algorithms

JEFFREY L. MARSH

As with most aspects of cleft care, management of velopharyngeal dysfunction (VPD) requires choices among different treatment modalities, technical variations within a specific modality, and timing of intervention. I have consolidated my experience, as a

member of an interdisciplinary team managing VPD over the past 26 years, into a series of algorithms (Figs. 35.1–35.4). These algorithms not only guide my personal patient care decision making but have proven useful in teaching others about VPD manage-

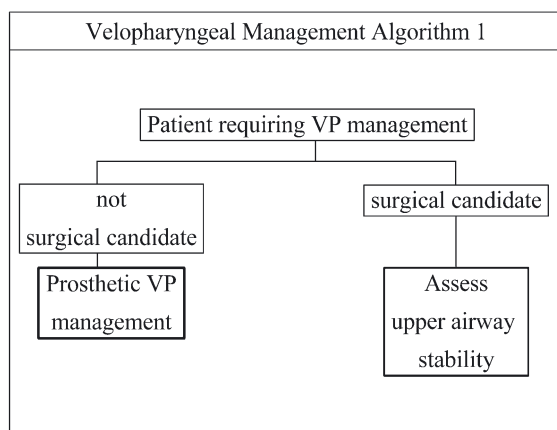


Fig. 35.1. Velopharyngeal management algorithm 1

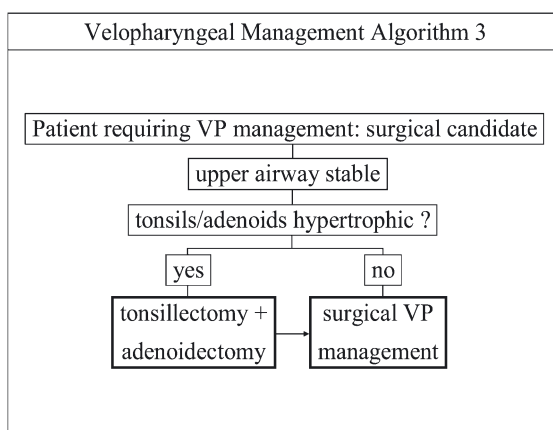


Fig. 35.3. Velopharyngeal management algorithm 3

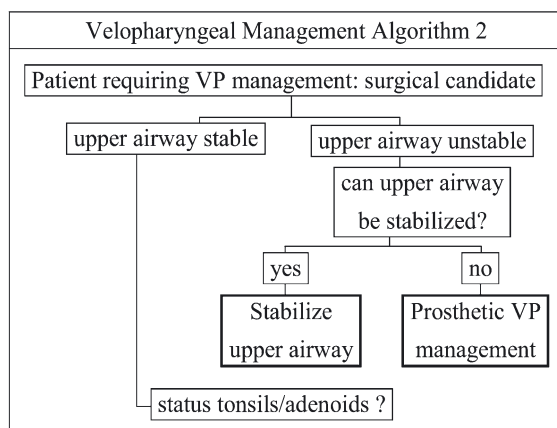


Fig. 35.2. Velopharyngeal management algorithm 2

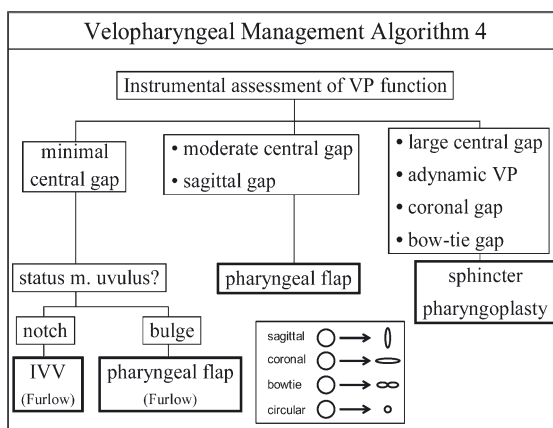


Fig. 35.4. Velopharyngeal management algorithm 4

ment [20]. This chapter presents and discusses these algorithms.

Individuals with possible VPD present to individual members of the cleft palate team or to the team as a whole for diagnosis and treatment. Historically, the large majority of such individuals had a repaired cleft palate and a small minority, without overt cleft palate, developed VPD following adenoidectomy. While post-palatoplasty patients with impaired speech still comprise the largest group evaluated for VPD [10, 15, 24], individuals with velocardiofacial syndrome, neurological impairment, and postobstructive sleep apnea surgery (uveopalatoplasty) comprise a steadily increasing percentage. Whereas identification of the etiology of the VPD is critical to assigning differential management based on differential diagnosis [17, 19], the evaluation of velopharyngeal function is the same regardless of etiology [37]. I shall first discuss the evaluatory process and then specific managements.

35.1 Evaluation of Velopharyngeal Function

Speech results from modulation of the pressurized air stream that emerges from the lungs during expiration. A number of structures between, and including, the glottis and the lips (articulators) modify that air stream to produce specific sounds. One of those modifiers is the velopharynx, which normally is a dynamic sphincter. A sphincter is an anatomic structure that can open or close to varying degrees and speeds depending on the physiological necessity. The tissues that comprise the velopharyngeal sphincter are the velum (soft palate), the right and left lateral pharyngeal walls, and the posterior pharyngeal wall. The space enclosed by and modified by the velopharyngeal sphincter is the velopharynx. The velopharynx is the connection between the nasopharynx and the oropharynx. Normally at rest it is a wide opening through which air flows easily and secretions drain; when contracted, it closes the nasopharynx from the oropharynx preventing passage of gases, fluids, and solids between the two cavities. Dysfunction of the velopharyngeal sphincter impairs that separation of the nasal and oral cavities for both deglutition and speech to varying degrees. Outcome assessment of VPD management requires consideration of velopharyngeal function for all of the following physiological activities: speech, breathing, swallowing, handling nasal secretions, and sleep.

There is a lack of consensus on the preferred term to describe such dysfunction: *velopharyngeal incompetency*, *velopharyngeal insufficiency*, and *velopharyngeal inadequacy* have all been abbreviated as *VPI*. I prefer the term *velopharyngeal dysfunction*, abbrevi-

ated as *VPD*, since it connotes physiological impairment without attempting to denote etiology [8, 16]. Regardless of the terminology, a specific set of physical findings suggest VPD. These include excessive nasal resonance during speech (hypernasality), inappropriate nasal air-flow noise (nasal turbulence), and abnormal facial movements (facial grimacing). An individual with one or any combination of these findings will be referred for evaluation of possible VPD.

The first task for the VPD team is to determine whether the patient actually has VPD. This determination has traditionally been made solely on the basis of a perceptual speech/language evaluation by a speech/language pathologist with specific training and experience in velopharyngeal function [41]. This evaluation includes both spontaneous speech and a provocative speech sample, structured to specifically challenge the velopharyngeal mechanism with increasingly more complex tasks [24, 25]. The primary activity of this perceptual assessment is auditory, that is listening for the production of specific phonemes. However, observing the face for grimacing, watching for fogging of a mirror below the nares, and feeling for airflow through the nares with attempted pronunciation of phonemes that require velopharyngeal function are other perceptual assessments that assist the screening evaluation. Some professionals now argue that instrumental assessments of oral-nasal airflow should be part of this determination process. However, we have not yet incorporated nasalance measurement into our initial assessment.

The auditory perceptual screening of velopharyngeal function sorts the patient into one of three groups: normal VP function; minimal VPD without impairment of communication or psychosocial stigmatization; or sufficient VPD to interfere with speech comprehension by others and/or stigmatize the speaker. If the assessed individual screens normally, no further evaluation of the velopharynx is indicated until the return for routine follow-up or some concern regarding speech arises. If the possible dysfunction is minimal, meaning that the affected individual, and his or her parents in the case of a child, were not aware of a speech problem and the Cleft Team, in concert, feels that the dysfunction is unlikely to cause impairment of communication or have psychosocial consequences, then the affected individual will have a repeat auditory perceptual speech evaluation at an interval shorter than the usual follow-up and the individual/parents will be informed of the concern regarding velopharyngeal function as well as of the signs and symptoms of increasing dysfunction so that they can contact the Cleft Team should they occur. If, on the other hand, it is the perception of the sophisticated observer that there is a possibility of

morbid velopharyngeal dysfunction, further evaluation is considered.

If one accepts the principle that treatment should be diagnosis specific, and I do, then effective management of velopharyngeal dysfunction requires both recognition of the condition and definition of the responsible pathoetiology [41]. While auditory perceptual evaluation can separate normal from abnormal velopharyngeal function, it cannot identify the etiology of the dysfunction. Instrumental velopharyngeal evaluations, in contrast, can be etiology specific [12, 31, 40]. (Instrumental evaluations can be divided into those that visualize the velopharynx and those that indirectly assess its function.) [1, 5, 6, 10]. Clinically useful visualization utilizes still (lateral skull x-ray) or real-time x-ray (speech videofluoroscopy). Documenting the effect of velopharyngeal sphincter function on air flow air pressure or sound or light transmission across the velopharyngeal port produces indirect data about velopharyngeal function. A multidisciplinary consensus regarding reporting of visualization studies provides a useful means for standardized interpretation of such studies [9]. For the past 20 years, the cleft teams that I have been associated with have used both nasoendoscopic and fluoroscopic imaging of the velopharynx during standardized speech tasks to evaluate velopharyngeal function [4, 31, 32, 34]. These evaluations are audio-video recorded, previously on magnetic tape and now digitally, in order to:

1. Permit review by other members of the Cleft Team who were not present during the actual examination
2. Permit comparison of serial assessments of the same individual
3. Permit comparative assessment of the effects of therapeutic intervention in the same individual, and
4. Facilitate cross-sectional comparison of various interventions

These tapes are then reviewed by a subset of the Cleft Palate team, which consists of representatives from the disciplines of Otolaryngology, Plastic Surgery, and Speech/Language Pathology [3, 30]. A consensus opinion regarding interpretation of the images and preferred method of management is then arrived at by group discussion. The conclusions are communicated to the patient/family for reaction and to appropriate primary and secondary care providers, such as the patient's speech therapist and pediatrician/physician. When velopharyngeal management, either prosthetic or surgical, is recommended and performed,

the velopharyngeal function is re-evaluated 3 months after the intervention with auditory perceptual and instrumental evaluations mirroring those performed preoperatively.

35.2 Differential Diagnosis of Velopharyngeal Dysfunction

For the purposes of differential management based upon differential diagnosis, velopharyngeal dysfunction can be divided into four broad etiologic categories:

1. Anatomic deficiency
2. Myoneural deficiency
3. Anatomic and myoneural deficiency
4. Neither anatomic nor myoneural deficiency

Each category has several specific causes for that deficiency. *Anatomic deficiency* includes those static tissue problems that prevent adequate velopharyngeal sphincteric closure, such as an unrepaired cleft palate, a palatal fistula, a short velum, or a deep velopharynx. *Myoneural deficiency* implies that there is adequate palatal and velopharyngeal structural anatomy but one or more components of the velopharyngeal port have inadequate or absent function, such as may occur with head trauma or degenerative neurologic disorders. Combined *anatomic and myoneural deficiency* may occur with repaired cleft palate, unoperated submucous cleft palate, or postablative surgery and/or radiotherapy for oro-naso-pharyngeal malignancy. Rarely an individual presents who has *intact velopharyngeal anatomic structures and is myoneurally intact* but whose speech nonetheless exhibits signs and symptoms of velopharyngeal dysfunction due to either learned behavior, such as having a parent with cleft VPD and living in a rural area, or psychiatric disorder.

The initial goal of the instrumental evaluation of the presumably impaired velopharynx is to determine which of the four categories the patient belongs to since that broadly determines the type of therapeutic intervention(s) to be considered (see below). The combination of visual direct and indirect (mirror, endoscope) intraoral inspection and indirect velopharyngeal functional inspection (nasendoscopy, multiview fluoroscopy) permit definition of the structural and functional impairments contributing to the VPD. Once the category has been defined further general health history, physical examination, and testing determines the specific etiology for that patient's VPD.

35.3 Differential Management of Velopharyngeal Dysfunction

The four etiologic categories enumerated above broadly determine the differential management of velopharyngeal dysfunction. *Anatomic deficiency* is usually managed by restoration of the deficiency by either surgical or prosthetic means [20] (Fig. 35.1). *Myoneural deficiency* is usually managed prosthetically until the neurologic status has stabilized and the patient becomes a candidate for velopharyngeal surgery, which may or may not occur [26]. *Combined anatomic and myoneural deficiency* is usually managed by restoration of the deficiency by either surgical or prosthetic means. Speech therapy, often pre- and usually postintervention, is an essential element of management of these three categories in addition to surgical or prosthetic intervention. For *learned or behavioral VPD*, speech therapy with or without behavioral management is the treatment of choice; there is no role for surgery in such cases [11].

Prosthetic management for VPD is indicated when:

1. The patient is unsuitable for surgical velopharyngeal management due to other major medical problems, such as inoperable congenital cardiac disease or severe pulmonary insufficiency (if and when the other medical problem becomes resolved or stabilized enough to minimize the anesthetic/surgical risk, conversion to surgical velopharyngeal management is considered);
2. The parents/legal guardians refuse to permit surgery because of religious or emotional considerations;
3. The patient has a complex speech/language problem and it is not clear that velopharyngeal management will significantly improve speech intelligibility (if the trial of a speech prosthesis demonstrates significant improvement in intelligibility, then conversion to surgical velopharyngeal management is considered);
4. The patient has a neurological condition that includes VPD and it is not clear whether the neurological deficit has stabilized and/or that velopharyngeal management will significantly improve speech intelligibility (if the trial of a speech prosthesis demonstrates significant improvement in intelligibility, and the neurological status has stabilized, and the patient is not a significant anesthetic risk due to the neurological pathology, then conversion to surgical velopharyngeal management is considered);
5. The patient has an unstable upper airway that cannot be stabilized (Fig. 35.2) and VPD (this was the major indication for prosthetic velopharyngeal management at our Cleft Center prior to the introduction of mandibular distraction osteogenesis for

stabilization of the upper airway in individuals with mandibular microretrognathia).

When a prosthesis is used to manage VPD, the type of prosthesis depends on the amount and quality of palatal tissue and the findings on nasoendoscopic velopharyngeal functional assessment [35, 38]:

1. When there is adequate velar tissue (a long, unscarred velum), a palatal lift is prescribed;
2. When the velar tissue is deficient (a short, scarred velum with a deep pharynx), a velopharyngeal obturator is prescribed;
3. When the velum is long but the pharynx is deep or a palatal lift fails to achieve complete velopharyngeal closure, a combined lift and obturator, "liftortor," is prescribed.

Surgical VP management is recommended for all other individuals with VPD who do not meet one of the criterion listed above for prosthetic management. Several types of surgical alteration of components of the velopharynx are possible:

1. Velar lengthening (palatal pushback, Furlow double opposing Z veloplasty);
2. Intravelar muscular reconstruction;
3. Pharyngoplasties (pharyngeal flap, sphincter pharyngoplasty);
4. Posterior pharyngeal wall augmentation.

Each type of surgical alteration of velopharyngeal structures has its advocates. Although some comparisons of two or more methods of VPD management have been reported, they fail to meet rigorous scientific criteria for discriminating among therapeutic options [7, 22, 29, 33, 39].

Both types of velar operations and posterior wall augmentation procedures are fundamentally different from, and theoretically more physiologic than, the pharyngoplasties in that they do not create a permanent subtotal obstruction of the velopharyngeal port. Nasal physiology is less likely to be impaired with velar and posterior pharyngeal wall procedures than with pharyngoplasties. The challenge for the velopharyngeal surgeon is to provide the patient with the means to achieve velopharyngeal closure without obstructing the nasal airway for breathing, secretion control and passage of air into the nose during speech for those sounds that require nasal resonance. For this reason, when either a pharyngeal flap or a sphincter pharyngoplasty is elected, a tonsillectomy and adenoidectomy are performed at least 3 months prior to the velopharyngeal procedure so that hypertrophic lymphoid tissue cannot obstruct the surgically diminished velopharyngeal port(s) and to facilitate the performance of the pharyngoplasty.

So how does the surgeon choose among these options for intervention? In collaboration with my colleagues on the multidisciplinary velopharyngeal team, I choose among the therapeutic options based upon the anatomy and function of the velopharynx as documented by nasoendoscopic and multiview fluoroscopic visualization of the velopharynx [2], performing both spontaneous speech and standardized speech tasks. The selection process follows an algorithm that has been in use, with minor modification, over the past 14 years (Fig. 35.4). My rationale for pairing of the residual velopharyngeal gap on attempted complete velopharyngeal closure with specific interventions follows.

35.3.1 Small Midline Gap

Management of a small midline gap on attempted complete VP closure depends on the status of the intravelar musculature. If the patient has not had an intravelar veloplasty or did not have a “radical” (method of either Sommerlad or Cutting) intravelar veloplasty, a radical IVV is recommended. The patient/family is informed that there is about an 80% change of correction of the VPD with such an intervention, in my experience, with no expected airway morbidity. They are also informed that either a narrow, lined, superiorly based pharyngeal flap or a sphincter pharyngoplasty could be performed with an almost 100% correction of VPD but with some risk of impairment of the upper airway, primarily intranasal retention of nasal secretions, and to a lesser degree nasal airway obstruction or the rare case of obstructive sleep apnea. If the radical IVV fails to sufficiently correct the VPD, then a pharyngeal flap or sphincter pharyngoplasty are offered with the caveats listed above. Some families prefer to avoid nasal obstruction even if there is a 20% chance of needing a second operation while others prefer more certainty of resolution of the VPD regardless of airway compromise. (Rearrangement of the velar soft tissues including the muscle via the Furlow double opposing Z-plasty is an alternative to radical IVV for the patient with a small midline gap on attempted complete VP closure. I am unaware of a prospective comparison of the efficacy and morbidity of these two approaches for equivalent VPD situations. The choice then between the two operations currently is surgeon’s preference. I have no personal experience with the Furlow but several of my associates, current and past, use it for this indication as well as several cleft surgeons whose technical skill and reporting honesty I respect.)

35.3.2 Sagittal Gap with Active to Moderately Active Lateral Pharyngeal Wall Motion

I reserve the pharyngeal flap operation for VPD due to a sagittal gap with active to moderately active lateral pharyngeal wall motion. Because of this restriction, I only perform narrow or moderately wide, superiorly based, lined pharyngeal flaps. (I stopped performing wide obstructive pharyngeal flaps in 1989, when sphincter pharyngoplasty entered our therapeutic repertoire, due to the consistent morbidity of wide obstructive pharyngeal flaps with respect to the upper airway: intranasal retention of nasal secretions, obstruction of the nasal airway with obligatory mouth breathing, hyponasal resonance, and obstructive sleep apnea as well as death [14].) A review of 71 patients, who underwent a pharyngeal flap operation at our cleft center between 1982 and 2000 and had adequate preoperative and 3-month postoperative perceptual and instrumental velopharyngeal speech function evaluations and a 12-month postoperative perceptual velopharyngeal speech evaluation, was conducted in 2001. This study documented satisfactory nasal resonance in 74% of patients so selected and treated. Of the remainder, surgical tightening of the one or both lateral ports with incomplete closure, as documented on postpharyngeal flap nasendoscopy, increased the resolution of symptomatic VPD to 92%. Postoperative complications included obstructive sleep apnea in 5 patients, symptomatic nasal secretions in 7 patients and obligate mouth breathing in 5 patients. Denasality (hyponasal nasal resonance) was noted in 21% of patients at 3 months postoperatively but had diminished to only 6% at 12 months postoperatively. Of the patients receiving a pharyngeal flap, 3/4 had cleft palate +/- cleft lip, 1/4 were noncleft VPD, 1/5 of the patients were syndromic. Postoperative nasal resonance was not affected by the presence or absence of cleft palate ($p = 0.7$) or the presence or absence of a syndrome ($p = 0.2$) [28].

35.3.3 Hypodynamic or Adynamic Velopharyngeal Sphincter

I utilize the sphincter pharyngoplasty (Jackson modification of the Hynes procedure with “crossed-arms” overlapping posterior tonsillar pillar myomucosal flaps) for all cases of VPD secondary to a hypodynamic or adynamic velopharyngeal sphincter [38]. Preoperative specification of the locus for insertion of the myomucosal flaps on the posterior pharyngeal wall is based on the level of maximum attempted velopharyngeal closure, as documented on the lateral fluoroscopic VP evaluation [27]. A review of our sphincter

pharyngoplasties in 2000 [21] documented resolution of symptomatic VPD in 72% of 162 patients so selected and treated. Of the remainder, surgical tightening or reconstruction of the central port with incomplete closure, as documented on postpharyngeal flap nasendoscopy, increased the resolution of symptomatic VPD to 85%. For the 11 residually symptomatic patients, a third tightening was performed or a narrow pharyngeal flap was placed in the center of the residual sphincter pharyngoplasty port. Significant hyponasality was noted in only 10% of patients.

35.4 Conclusion

Differential diagnosis of velopharyngeal dysfunction, using instrumental visualization of velopharyngeal function, allows for differential therapeutic management [23, 29]. The objective of such management is to optimize the function of the velopharynx for speech tasks while minimizing the morbidity of the intervention upon the upper airway. My experience with such an approach over the past 21 years validates the assumption that differential management of velopharyngeal dysfunction based upon differential diagnosis can achieve this goal.

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36 Optimal Age for Palatoplasty to Facilitate Normal Speech Development: What is the Evidence?

SALLY PETERSON-FALZONE

Surgeons, speech pathologists and other members of cleft palate/craniofacial teams have been trying for decades to gain some kind of “sure grip” on the relationship between age at palatoplasty and the child’s speech development. I reviewed the literature on this topic in 1996 [65], and extended that review considerably in 2001 in concert with my coauthors of the third edition of *Cleft Palate Speech* [66]. Both reviews pointed out the differences between unverified clinical commentary (e.g., “Our speech results were good.”) and scientific proof of investigators’ conclusions about speech and language development. Much of the literature consisted of undocumented clinical insight, although there were many data-based reports as well. The reliance upon clinical insight and upon case reports that lack independent documentation of results still prevails as of this writing. On the one hand, one must commend the continuing and, indeed, zealous pursuit of this critical question. On the other hand, we must ask if there is a way to increase the relative proportion of reliable, valid data and decrease the dependence upon undocumented opinion.

36.1 Why Is It So Difficult to Answer this Question?

Clinical research is inherently hazardous because so many factors affect the outcome of a study. The investigator(s) *cannot* control all the factors the way they would in a laboratory study. Far too often, however, the professionals who have tried to determine the optimum timing for closure of a cleft palate have not fully recognized the possible contaminating factors, such as:

1. Patients with varying types and extents of clefts, heterogeneous socioeconomic backgrounds, varying health status and health-care history (adequacy of pediatric care, otologic history and documentation of hearing status, adequacy of early feeding and growth, adequacy of stimulation in the home);
2. Multiple surgeons *presumably* performing the same surgical procedure;
3. Children undergoing the same physical procedure but at different times in their chronologic *and* developmental ages;
4. Children whose pre-and postpalatoplasty care may or may not have included regular follow-up evaluations by the cleft palate/craniofacial team (see recommendations from the American Cleft Palate-Craniofacial Association, 2000);
5. Children whose pre-and postpalatoplasty interventions may or may not have included early childhood stimulation programs;
6. Highly variable documentation of the status of velopharyngeal closure postpalatoplasty. The substance of postoperative evaluation continues to vary widely. The low end of the spectrum consists of non-data-based statements such as “the palate appeared to move well after surgery” or “speech was significantly improved.” The upper end consists of well-documented objective measures in combination with perceptual assessment of speech.
7. One-time postoperative assessment versus longitudinal data (kids grow, adenoids usually go away, maturity eventually kicks in, etc.);
8. Children who have undergone primary veloplasty (at varying ages), with inconsistent use of interim obturating plates (for varying lengths of time, and typically with very little documentation of that use) and later surgical closure of the hard palate (again, at ages that vary from less than 1 year to the early teenage years)¹.

¹ The topic of primary veloplasty is a particularly difficult one to pursue in any scientific endeavor to answer the question of how early (or late) definitive palatal closure should be accomplished, and will therefore be discussed in Sec. 36.6.

Thus, the clinician who is trying to gather information on this topic must examine *each* report for the flaws that may have led to false conclusions.

36.2 Why Do Speech-language Pathologists Worry So Much About Early Development?

One concept that may seem foreign to practitioners whose primary focus is the physical status of the child's palate, teeth, or oropharyngeal mechanism is that what an adult hears as normal speech (or speech development) in a toddler is *not* simply the result of that toddler echoing what he hears around him. Normal communication development depends upon many factors: normal hearing, normal oral and pharyngeal structures, adequate stimulation from the environment, and reinforcement from that environment for communication efforts. A child who experiences communication failure on a consistent basis will simply give up (unconsciously), and settle for the simplest signals he can produce (grunts, whines, cries, etc.) in order to make the world pay attention. Try to envision an otherwise normally-developing child at the age of 1 year who is walking (or nearly ready to do so), smiling, and vocalizing but whose first attempts at words consist of only nasal consonants, grunts, and vowels. "Mama" will be produced normally (the consonant /m/ does not require velopharyngeal closure), but any attempt at "Dada" or "baba" (bottle) will sound like "uh uh." By contrast, a 12-month old who can produce "b," "d," "g," plus some of the consonants that do not require velopharyngeal closure (the nasal consonants, /h/, /w/, and "y") will probably be producing 10 recognizable (if simplistic) words. (Please see Fig. 36.1). His parents will reward these efforts with smiles and expanded input, e.g., "Da?! Do you want Daddy?" The

child who cannot produce these speech patterns at this very young age will quickly recognize that his communication attempts are useless. He cannot convey much about what he wants, needs, or feels, and will conclude that those around him do not care, "so why bother?" These early failed communication attempts have a long-term impact not only on language development but the child's overall cognitive and psychoeducational development. Communication success vs. communication failure at 12–18 months of age will inevitably become either wonderful or disastrous well before the age of 3 years. The potentially dismal long-term outlook for the child's well-being thus becomes the concern of all his care-givers, including all members of the cleft palate/craniofacial team.

36.3 What Constitutes Scientific Evidence in Clinical Research?

The strongest evidence that a particular treatment protocol is better than another comes from prospective clinical trials that include study not only of the "treatment group" but a carefully matched control group over the same period of time [2, 22]. Prospective studies with a study group and a control group are also termed "cohort studies" [78]. This is in contrast to "case control studies," which are usually retrospective in that they compare current cases with controls from the past (perhaps consisting of untreated cases or cases treated by an older method). Prospective clinical trials are the most difficult to execute. In fact, Wissow and Pascoe [78] warned

"Clinical trials are poorly suited to studies of (1) multiple therapeutic modalities (because too many subjects are needed to evaluate the many possible therapeutic combinations); (2) small changes in a therapeutic plan

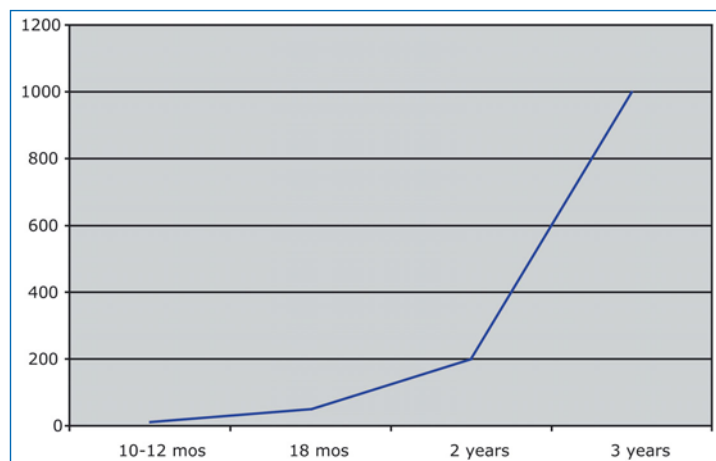


Fig. 36.1. Vocabulary development within the first 3 years of life. The number of words used at 10–12 months of age is not "0" but usually 2–10 words. By 18 months, most children have at least 50 words, and in the next 6 months that number quadruples to 200. By the age of 3 years, most normal children have speaking vocabularies of about 1000 words.

(the effort it takes to do the study may outweigh the potential significance of the outcome); (3) therapies that may be changed during the course of the study so that the results are at risk for becoming obsolete before the study is completed; and (4) treatments with only rare outcomes or outcomes that will be observable at a time far distant in the future.”

Note how strongly numbers 1, 2, 3, and the second half of 4 pertain to the study of the best timing for palatoplasty.

Shprintzen [71] made several observations that should be kept in mind by anyone engaged in clinical research, and certainly by those who would study the question of the optimum timing of palatoplasty. He pointed out that most research is guided by a specific motive, usually a vested interest of the investigator, meaning the individual has an *a priori* bias regarding the outcome. With regard to sample selection, he pointed out the necessity of acknowledging the heterogeneous etiology of clefting, and the influence this variation may have on development. He stated (p. 137), “...if a unilateral cleft lip and palate occurs because of an intrinsic hypoplasia of one maxillary process, it would be anticipated that the intrinsic hypoplasia would remain a factor throughout postnatal growth...If a major source of variability [in postsurgical growth] is intrinsic differences in facial growth potential related to population heterogeneity, then the subject population must be made as homogeneous as possible.” He observed that, while investigators may have tried to eliminate all syndromic cases in their studies of this issue, none of the studies published up to the date of his commentary had held constant all the variables that could influence treatment outcome. At the same time, he warned (p. 138), “Holding all variables constant in patients with multiple problems is not only difficult, but the ethics of withholding treatment for the purposes of determining research outcome might have some difficulty passing an Institutional Review Board.”

Table 36.1 rank-orders research designs by the strength of the evidence they can provide. In a simpler form, we may segregate the aspects of research design that lead to stronger supportive evidence for a particular treatment approach, versus those that weaken a study's conclusions, into “undesirable” vs. “desirable”:

“Undesirable”	“Desirable”
Investigator undertakes the study to prove that his treatment protocol is better than another	Investigator has no prior bias as to outcome of the study
Results recorded only by the investigator without independent observations by unbiased judges	Unbiased observers/ recorders of results
Single recorder/observer	Multiple independent observers/recorders, with measurement of intra- and interobserver reliability
Longitudinal observation of results	One-time assessment of results
Retrospective review of records	Prospective study
Single group of patients, treated according to one protocol, no control group	Cohort studies, comparing two or more groups of homogeneous patients, one treated according to the protocol under study, the other untreated or treated by a standard accepted protocol
No attempt to control for the multiple variables that may influence results	Control, or at least careful independent documentation, of all variables (e.g., absence of associated anomalies, type and extent of cleft, health history, ear disease and hearing loss, socioeconomic factors including parental nurturing and stimulation, etc.)

Table 36.1. Levels of evidence in clinical practice, ranked from weakest to strongest, and what each type of evidence would require in a study attempting to relate age of palatoplasty to speech outcome. Designs “B” through “F” would require IRB (Institutional Review Board) review. Please see comments regarding HIPAA (the Health Insurance and Accountability Act of 1996), following this table

Source of evidence	What is involved	What would add strength to the evidence	Comments regarding the value of this form of evidence
A. “Expert opinion” or “clinical insight”	Minimally, one person’s opinion on what works best	Agreement by other “experts,” each with appropriate experience	Relatively easy to find; most vulnerable to dispute
*B. Retrospective case studies; all information taken from archived records (with or without historical controls)	Independent non-biased personnel who examine a pre-determined set of data recorded in a uniform format	Demonstration of intra- and interclinician reliability; historical controls would be patients with the same types and extents of clefts, non-syndromic, treated by some other surgical protocol	Probably the most common source of data on this question; subject to bias, especially if one or more of the investigators has a predetermined opinion; most difficult aspect is controlling for the myriad independent variables (e.g., hearing loss, overall health, socioeconomic factors, etc.) that affect outcome
*C. Retrospective case studies (archived information) combined with current clinical assessments	As in (B), above, plus current assessment of outcomes by independent, non-biased personnel using the same assessment protocol with all patients	Demonstration of intra- and interclinician reliability; control group(s) treated by other surgical protocols	Case histories + clinical assessments are very common, but, as in (B) above, investigator bias is frequently built into the research and control of independent variables is difficult
*D. Cohort studies (retrospective)	Investigator pairs each patient treated by Method X with a patient treated by Method Y. Each of the pair must match for the independent variables that could influence outcome	Demonstration of intra- and interclinician reliability; the larger the number of patients in each treatment group, and the longer the period of observation, the stronger the evidence	Currently a favored approach in studying this question, but extremely difficult to control for all the independent variables
*E. Cohort studies (prospective)	Investigator establishes the matched groups of patients before any treatment is rendered	Strength of study depends upon the investigator’s ability to hold everything constant for each pair in the matched groups except for outcome; longitudinal follow-up of outcome strengthens the study	Very difficult to execute due to demands of matching patients on all independent variables; also, in longitudinal studies, patient populations change over time (growth, health factors, relocation, etc.)
*F. Prospective randomized trials	Investigator randomly assigns treatment protocols (i.e., X, Y, Z) to patients who are matched for type and extent of cleft, age, sex, other health factors (ear disease, etc.)	As in (E), above	Randomizing treatments raises serious ethical issues; patient consent is a must. Hospital IRB approval difficult to obtain. Strongest level of evidence, but rarely achieved

* The HIPAA (Health Insurance and Accountability Act of 1996) prohibits research personnel in health care facilities from combining patient records unless permission has been granted by the patients themselves. The only person who can recruit the patients is the physician (clinician) who provided the care to the patient. This has an obvious impact in cleft palate/craniofacial care, where the preferred approach to care is through a multidisciplinary team (American Cleft Palate-Craniofacial Association 2000), and not by a single health-care professional. Historically, permission for examination of records or the sharing of information from patient records has not usually been obtained in the form currently required by HIPAA, rendering most retrospective studies impossible if they must meet HIPAA requirements. Information on HIPAA is available on the web at <http://www.hhs.gov/ocr/hipaa/>.

The summary to both Table 36.1 and the comments above might be “If you want to prove that a given surgical protocol produces better speech results in non-syndromic children with clefts, you need to plan a prospective, randomized study with matched controls; two or more surgeons performing the same procedure (to prove that it is not the surgeon’s technique that is making the difference); *all* independent variables matched across patient pairs; large numbers of patients in each treatment group; results recorded by independent observers on standardized forms (not just anecdotal comments) and recorded over time rather than in a single posttreatment observation.”

Obviously, this description is one of a utopia that one can only keep in vision, not plan on reaching. The clinical researcher is therefore charged with the task of trying to come as close to this as possible, and being very careful in the interpretation of the results, taking into account all the factors that could not be controlled.

36.4 A General Warning: “A” Does Not Cause “B”

Because we clinicians are so anxious to answer the question that is the focus of this chapter, we are very prone to jump to conclusions about cause and effect. The most common trap is assuming that because “B” is strongly associated with “A,” even if “B” *always* follows “A,” that “A” *causes* “B.” Friedman et al. [22] termed this “causal inference.” A widely used analogy points out that Tuesday always follows Monday, but does that mean Monday causes Tuesday? Even if we were to see that maxillary arch collapse is present in 100% of children with cleft lip and palate following palatoplasty at 12 months of age, that does *not* mean that either the palatoplasty technique or the age at palatoplasty caused the arch collapse².

Cleft palate is not just a rearrangement or “disarrangement” of structures, but an inherent deficiency of tissue. The mere performance of palatoplasty, the particular surgical procedure employed, or the age at which it is performed thus cannot be blamed for subsequent arch collapse. On the other side of the argument, none of these three factors, alone or in concert can be held responsible for either normal or abnormal subsequent development of communication development. If we were to segregate the physical vari-

ables from the nonphysical variables that determine outcome from palatoplasty, we would be looking at the following:

Physical factors	Nonphysical factors
Type and extent of original cleft	Developmental level (chronological age compared to cognitive and communication development)
Presence/absence of associated anomalies; syndromes, etc.	Parent-infant bonding; parental stimulation
Overall health history (particularly with regard to infant feeding and otologic health)	Other psychosocial issues affecting the family
Adequacy of pediatric care both before and after surgery	Adequacy of relationship between surgeon (or team) and the family both before and after surgery

36.5 A Review of the Literature up to 2000

The review in 2001 [66] could be current only up to a few months before publication of the book. The main points of information were as follows:

1. There was a very wide range in what clinicians considered to be “early” palatal surgery, e.g., the first 28 days of life [15] 9–25 weeks [9] up to 2–3 years [42].
2. Judgment as to whether or not palatoplasty had in fact provided an intact palate often consisted of only visual observation of the oral cavity³ (and sometimes only on a one-time basis), sometimes combined with rather pathetic “objective” measures, e.g., fogging on a mirror held beneath the child’s nose during speech [5]. At the other end of the continuum, many studies combined rather rigorous perceptual data with a full armamentarium of instrumental assessments.

2 Shprintzen [71] made this same point: “Basing cause and effect relationships on the correlation of two factors is scientifically unsound, yet is undoubtedly the most common error made in research design and interpretation.”

3 The examiner *may* be able to tell from the intraoral view alone whether or not the repaired palate is intact, but the velopharyngeal system is *not* visible because VP closure is the closure of the upper (nasal) surface of the velum against the posterior pharyngeal wall. Visualization of VP closure requires radiographic (usually fluoroscopic) or endoscopic instrumentation. In the future, magnetic resonance imaging may become a reasonable alternative.

3. The increased knowledge we gained in the 1980s and 1990s regarding the effects of early structural constraints on prespeech vocalizations and on later speech development in children with clefts could constitute a very good reason to perform palatal closure early, e.g., before the child reaches the milestone of producing his first meaningful words [7, 8, 19, 26, 27, 62, 63]. Chapman [6] found, as had previous researchers, that the process of phonologic acquisition in youngsters with clefts was "...slower, perhaps due to lingering or past articulatory and/or structural constraints on the speech mechanism."
4. However, a previous claim that the chronological age of 12 months for completion of palatal closure was a cut-off line for preventing the development of compensatory articulations in children with clefts [12, 13] was not supported by subsequent studies [11, 64] in which an attempt was made to verify that cut-off age.
5. Despite the fact that their results could not be replicated in subsequent studies, Dorf and Curtin [12] had made one very important point that was largely ignored by other investigators, namely that the child's "articulation age" (level of phonologic and phonemic development), *not* his chronologic age, that should be a prime consideration when trying to plan the best timing for surgery. That is, speech and language development vary among children: Some are attempting to produce 10–12 meaningful words at that age, while others are just beginning to use "mama." The more sounds and words the child is trying to produce, the more urgent the need for an intact palate and velopharyngeal system.
6. Clinical surveys in the 1960s, 1970s, 1980s, and 1990s awakened us to the fact that associated anomalies, sequences, associations, and syndromes were probably affecting development in at least 50% of children with clefts [39, 72, 73, 80]. It is very easy for anyone other than an experienced dysmorphologist to miss subtle signs of other congenital defects in children with clefts. Furthermore, it is easy to miss signs of cognitive delays, hearing loss, and other threats to development, particularly if a child is *not* evaluated by an interdisciplinary team. Mixing surgical results from normally developing children with results from syndromic, multiply involved, or cognitively delayed children seriously impairs our ability to make decisions about the effects of any specific treatment.
7. Studies published during the 1990s on the ostensible relationship between timing of palatal surgery and subsequent speech development tended to support performance of surgery in the first 12 to 18 months of life, the exception to this general trend being the studies on primary veloplasty (reviewed later in Sect. 36.6).
 - a. Although their report was entitled "Correlation between the age at repair and speech outcomes in patients with isolated cleft palate," Haapanen and Rantala [28] did little to clarify the age-at-surgery question because they had only small, uneven numbers of subjects; none were operated before the age of 1 year; and the speech results consisted only of general categorizations based on one-time perceptual judgments without independent or objective documentation. These authors reported better speech in children whose palates had been closed between the ages of 16 and 20 months, as opposed to those whose surgeries had taken place either earlier (12–15 months) or later (21–24 months). They came to this conclusion because none of the children in the middle group (16–20 months at closure) had developed compensatory articulations. However, the number of children in this group was less than half the number in each of the other two groups. This fact, plus the one-time-only assessment with no longitudinal data, renders the conclusion tenuous at best.
 - b. Marrinan et al. [57] segregated 228 patients with four types of clefts (soft palate only, clefts of the hard and soft palate, unilateral cleft lip and palate, bilateral cleft lip and palate) into four groups based on age at closure: 8–10 months, 11–13 months, 14–16 months, and over 16 months. They found that secondary management for postoperative velopharyngeal inadequacy was necessary in 11% of the 8–10 month group, 14% of the 11–13 month group, 19% of the 14–16 month group, and 32% of the 16+ month group. This linear relationship between age at palate repair and need for a pharyngeal flap was reported to be statistically significant at the $p = .025$ level. The likelihood of need for a pharyngeal flap was much greater in those children with clefts of the hard and soft palate (no lip cleft) or BCLP in comparison to those with clefts of the soft palate only or UCLP. In fact, in the latter two groups, age at palate repair was not statistically significant (ranging from 10% need for pharyngeal flaps in the earliest repair up to 18% in those repaired over the age of 16 months). The authors attributed this difference to the size of the cleft, since the vomer bone was attached in these two groups, where as the need for secondary surgery in children with an unattached vomer (complete clefts of hard and soft palate or bilateral clefts) ranged from 12% in the earliest repair group up to 50% in those repaired over the age of 16 months.
 - c. Ysunza et al. [82] compared speech results in one group of children ($n = 41$) whose clefts were closed at 12 months of age to another group

($n = 35$) who were operated on at 6 months. This was a multifaceted study in which the postoperative evaluations included standardized perceptual evaluation of speech, videofluoroscopy, and nasopharyngoscopy. Somewhat surprisingly, early phonologic development was significantly better in the second group than in the first. In addition, none of the 6-month group showed subsequent development of maladaptive compensatory articulations, even though 6/35 (17%) of them showed evidence of postoperative velopharyngeal inadequacy. By contrast, in the 8/41 (19%) of the 12-month group that had VPI, 5 of these (62%) developed compensatory articulations. There was no difference between the two groups in terms of degree of maxillary collapse when the patients were examined at 4 years of age. None had had either preoperative or postoperative orthopedic treatment. The authors concluded (p. 678), "Because maxillofacial growth was not significantly different in [the two] groups of patients, it seems that cleft palate closure at 6 months of age is a safe and reliable procedure for correcting velopharyngeal function in cleft palate patients."

It is interesting that Ysunza et al. [82], using a full range of perceptual and objective tests, found a significant difference between children whose palates were repaired at 6 months of age and those repaired at 12 months⁴. As pointed out in 2001 [66], these results may speak to the value of the theoretical framework proposed by Kemp-Fincham et al. in 1990. The latter authors, reviewing the literature on the development of speech motor control and phonetic development in infants, concluded that there is a particularly sensitive period or state of readiness between the ages of 4 and 6 months. Perhaps palatal closure before or during this time frame is important if we are to avoid the development of the maladaptive compensatory articulations that are so devastating to a child's early communication attempts [41].

d. A few surgeons in the 1990s were performing very early palatal closure (often simultaneous with lip closure) in the first weeks of life [9, 15]. However, there have been no data on speech in the babies so treated, nor has there been any independent substantiation of any of the early physical results of the surgery. At this point in time, caution seems warranted.

36.6 The Ongoing Dilemma of "Primary Veloplasty"

The 1980s and 1990s saw a large number of studies on two-stage closure of the palate, with soft palate closure (often combined with lip closure) taking place prior to closure of the hard palate. The aim of this approach is to protect craniofacial growth from the presumed negative effects of hard palate closure. In some treatment centers, obturating plates have been placed over the residual cleft during the interim between veloplasty and hard palate closure, although the use of such plates has been inconsistent between centers and across patients within the same center. The timing of both the primary veloplasty and the later closure of the hard palate has varied greatly among these studies. Age at soft palate closure has variously been the first few weeks of life, 3 months, 6–8 months, 12–18 months, and up to the late age of 2 years 6 months; timing of hard palate closure ran the gamut from 12 months up to an extreme of 11–13 years [10, 14, 17, 20, 21, 25, 30, 37, 46, 49, 50–53, 58, 60, 61, 67, 68, 70, 74, 76, 77, 81]⁵. In addition to the variability in timing and techniques of the eventual hard palate closure, study of the outcomes of primary veloplasty has been complicated by (1) use of infant orthopedics in some clinic populations, primarily in Europe (e.g., 24, 33–35, 83–85); and by (2) sporadic, inconsistent use of obturating plates while the hard palate remains open. Preoperative infant orthopedics⁶ is beyond the scope of this chapter, but it should be noted that there is

⁴ These authors were not the first to note a difference in speech outcome apparently related to differences in age at closure *within* the first year of life. Desai [16] and Copeland [9], in companion reports on the same groups of patients, reported differential effects of age at palatal surgery compared across 3 months, 4 months, 5 months, and 6 months of age. However, there were very uneven numbers of children in the four groups, and the short times separating the ages at surgery plus the rather questionable nature of the speech data (reportedly collected 5 years after palatal surgery but with an age range of the patients from "3.8 to 6.3 years") signaled the need for skepticism in interpreting the results.

⁵ In a very recent study by Lehner et al. [47], hard palate closure was performed first, between 4 and 5 months of age, with later closure of the soft palate at 10–17 months in children with unilateral clefts and "delayed" closure of the hard palate in bilateral clefts at 12–14 months. The authors concluded (p. 126), "There was no significant difference in terms of anterior and posterior maxillary width between early and delayed closure of [the] hard palate..." However, their timing of "delayed" hard palate closure was in no way comparable to what that term connotes in the rest of the literature.

⁶ It is very difficult to differentiate the information given in the literature about infant obturating plates vs. infant orthopedics. In the patient population of Hotz et al. [34, 35], the intraoral devices were meant both to mold palatal segments and to at least partially obturate the cleft.

little evidence that such devices either enhance pre-speech and early speech development through their effect on the movement of palatal segments, or adversely affect such development. There is a very old, as yet unresolved controversy as to whether or not such treatment has any beneficial effect, partially because "such treatment" is so variable in nature.

The story with regard to effects of obturating plates while the hard palate remains open is difficult to synthesize due to inconsistency of use. In the series of Lohmander-Agerskov studies, not all patients with open residual clefts were fitted with plates, and consistency of use by any single patient, by the authors' own acknowledgments, could not be fully documented. In 1998, Lohmander et al. concluded that the use of an intraoral plate to cover the residual cleft had not been proved to enhance articulatory development.

The data from Sweden [48–56] are by far the most extensive, and in some of their later work that team began carrying the initial palatal surgery (veloplasty) further forward, providing more extensive surgical closure of the palatal cleft and hoping to promote more postoperative spontaneous closure of the residual cleft. In each of the Lohmander-Agerskov studies, perceptual speech outcomes were rigorously documented. Overall, these studies leave us with at least the suggestion that, even with early speech intervention, the negative effects of delayed hard palate closure are revealed in aberrant speech articulation.

The overall conclusion in reading the Lohmander-Agerskov studies is that articulation was not as severely impaired as might have been expected in children who grew into their preschool and school-aged years with an open residual cleft, but that they exhibited hypernasal resonance, nasal air loss, and retracted articulation of dental and dento-alveolar consonants (meaning the use of abnormally-posterior placements for these consonants). Clearly, use of obturating plates did *not* prevent the development of retracted oral articulation, and in many cases this abnormal articulation persisted after palatal closure. It should be noted that very few of those children who exhibited abnormal articulation in these studies were using glottal placements (e.g., glottal stops), which are notoriously difficult to replace with appropriate oral articulation.

Of particular importance is a series of comments made by Dr. Lohmander-Agerskov and her colleagues with regard to effects of unrepaired palates on early speech development. In one of their early reports [49], they had no direct data on early speech development but stated (p. 30), "Presumably, some children with CLP develop misarticulations very early during babbling irrespective of early or late palatal repair." These authors said that, in recognition of this, their

"routines" had been changed to include "early intervention programs, evaluation of speech development, and early speech therapy if necessary." Later studies by the same group [50–52, 55, 56] confirmed that adverse effects of an open cleft were evident in babbling and continued to have an effect in speech sound acquisition in the toddler, preschool, and school-age years. In their 1998 paper, Lohmander-Agerskov et al. also noted a high prevalence of retracted oral articulation in 5-year-olds with large residual clefts, and the "marginal effect" of speech therapy on misarticulations.

Other clinicians studying effects of the primary veloplasty regimen reported poor speech results [3, 10, 37, 60, 68, 79]. Rohrich et al. [68] closed the hard palate simultaneously with the soft palate in 21 children at a mean age of 10.8 months (range 6 to 18 months). Another set of 23 children had primary veloplasty at a mean age 11.4 months (range 6 to 22 months) and hard palate closure at an average age of 48.6 months (range 30 to 57 months)⁷. They concluded (p. 236), "Our data suggest that delaying hard palate closure results in significant speech impairment without a beneficial maxillofacial growth response."

It is pertinent here to point out that some other studies on growth have also cast doubt on the effects of delayed hard palate closure on maxillofacial growth. Noverraz et al. [61] studied dental arch relationships in 88 children with unilateral clefts who underwent primary veloplasty at a mean age of 1.1 years, and who were segregated into four groups depending upon timing of hard palate closure (modified von Langenbeck in all cases): 1.5 years, 4.6 years, 9.4 years, and unclosed by the age of 10 years. All of these children had received presurgical orthopedic treatment according to the method of Hotz [33–35], but only until the time of soft palate closure. These authors found *no* differences in the dental arch relationships among the four groups. Thus, age at hard palate closure, at least within the range of 1.5 years up to 9.4 years, did not differentially affect maxillofacial growth. Gaggi et al. [23] examined maxillofacial growth in 30 individuals with unilateral cleft lip and palate whose palates had been closed in a single procedure between 11 and 14 months, compared to 30 who had had veloplasty between 18 and 24 months, and hard palate closure at 6 years. At 18 years of age, they reported a more severe impairment in growth of the maxilla in those who had

⁷ The large range of ages at surgery, both in the early complete closure and in the delayed closure groups, weakens the strength of this study.

had the two-stage repair⁸. Similarly, we should note findings of current studies regarding use of obturating plates in babies and toddlers with unrepaired clefts, since there may still be clinicians who think that the adverse effects of an open cleft on speech development can be avoided by use of a plate. Hardin-Jones and coauthors [29] studied 14 babies with unilateral and bilateral clefts, age 8 months to 15 months, who were fitted with palatal obturating plates and matched them to 14 babies of comparable ages who were not fitted. The authors found no differences between the groups in terms of the size of the consonant inventory or the place and manner of consonant production. In fact, there was a trend for the babies in the obturated group to produce more glottal consonants. The investigators concluded that palatal obturators do *not* appear to facilitate production of anterior consonants during babbling. Their conclusion seems to support the earlier observations of Lohmander-Agerskov and colleagues [55].

A much earlier report on “articulation development prosthesis” [18] yielded only limited data on this issue. These authors initially attempted to fit eleven toddlers with palatal plates. The plates were not built to completely close off the velopharyngeal deficit but only to obturate the oral portion of the cleft. The authors did not specify the age at which the appliances were placed. Only seven toddlers actually wore the prostheses; two of these seven did not return for follow-up, and three were still too young at the time the report was published for there to be any conclusions about speech outcome. Of the remaining two cases, one showed good articulatory development prior to palatal closure but the other developed compensatory articulations. Thus, the results were 50–50, no better than chance alone.

Konst and coauthors [43–45, 83–85] examined various aspects of communication development up to the age of 6 years in children with unilateral complete clefts who were fitted with orthopedic plates that partially covered their unrepaired clefts. This was a longitudinal study across three centers that were part of a consortium known as “Dutchcleft.” The treated group wore plates in the first year of life, up to the time of primary veloplasty at approximately 12 months of age. The first report [45] was on two groups of children, six fitted with the plates and six without. In the latest reports [43, 85], the number of children was still very small – just ten in each group.

The first article [45] examined prelexical development, meaning speech sound development before the first words appear. Although the treated group was using a greater proportion of alveolar articulations (meaning normal placements for alveolar consonants) at 12 months of age, the differences between the two groups was no longer apparent at 18 months. At the age of 2 1/2 years, the treated group was receiving higher scores in speech intelligibility than the nontreated group [83]. Between 2 and 3 years of age, the treated group was also using longer sentences than the nontreated group [44], but by 6 years of age the two groups presented with similar expressive language skills. The authors [44] concluded that treatment with intraoral orthopedics did not have long-lasting effects on language development.

However, in an expanded study published that same year, Konst et al. [84] asked trained listeners to rate thirteen specific speech characteristics and to indicate their total impression of speech in 10 treated children, 10 nontreated children, and 8 noncleft controls. The children at the time of the study were 2–3 years of age. The intelligibility rating scale was the single speech characteristic distinguishing the treated group from the nontreated group. The phonologic development was also closer to normal in the treated group. Both groups of cleft children differed from the noncleft group on nine of the speech scales, which is not particularly surprising since none of the children with clefts had had complete palatal repair. In a follow-up study on the cost-effectiveness of the infant orthopedic treatment, Konst et al. [43] reported that treatment produced an average improvement in speech of 1.34 points on a 10-point scale, at a cost of approximately 1041 Euro dollars. The concluded, “... from the perspective of speech development, the cost-effectiveness of [infant orthopedic plates] ... seems acceptable at this time.” The overall impression from this series of reports is that the authors found some beneficial effects of the plates on speech development, but the impact of their findings is somewhat weakened by the small number of children they were able to follow over time. Also, their data regarding cost consisted only of the direct cost of treatment, not the costs to families in terms of number of treatment visits, time off work, etc.

Van Demark [75] pointed out that advocates of primary veloplasty have ignored the financial and psychosocial costs of the increased number of surgical procedures (two palatal surgeries instead of one), clinic visits for the fitting and maintenance of plates, and the intensive speech therapy such as implemented in the Zurich treatment center [33–35, 76]. These remarks may have particular pertinence now in the USA, where third-party payers (insurance companies, HMOs) continue to cut services to the bare bones,

⁸ This study did not include data on speech, but one can only imagine the devastating effect on speech sound development in those children whose palates were left completely open until the age of 18–24 months. By 24 months of age, most children are using at least two-word sentences.

meaning that families who need more team visits and more surgeries are apt to be paying out of pocket.

As concluded 4 years ago [66], there is little doubt that those centers that have devoted so much effort to studying and improving treatment outcomes using primary veloplasty will continue to do so. However, speech-language pathologists remain skeptical, especially when the provision of a fully repaired palate during the crucial time for speech-sound acquisition can prevent the stigmata of “cleft palate speech.”

36.7 A Summary of What We Have Learned Since 2000

Recent studies providing some information relevant to the focus of this chapter may be summarized as follows:

1. Use of primary veloplasty continues. In the recently published study of Henkel et al. [32], speech results were determined *before* the hard palate had been closed, and the open cleft was not obturated⁹. It is thus not surprising that disordered articulation was present in half of the population of 24 children.
2. There is conflicting evidence regarding the possible benefit of obturating plates for promoting the development of appropriately-placed early speech sounds in children with unrepaired or incompletely repaired clefts. However, the largest amount of data on this approach comes from Sweden [50–52, 54, 55] and, combined with the study by Hardin-Jones et al. [29] casts doubt on the benefit of plates. The results reported by Konst et al. [43–45, 83–85] are pertinent, but the impact is weakened by the small number of subjects.
3. Some surgeons continue to pursue closure of palatal clefts in the neonatal period (first 28 days of life) [15, 69]. However, speech-language pathologists have no solid data indicating that these very early closures will benefit early speech development. To date, *no* follow-up data has been published on either speech development or craniofacial development in children operated so early, despite the fact that the first cases operated by Denk and Magee [15] must now be at least 9 years old.
4. Speech-language pathologists have continued to document the effects of an open cleft during early speech sound acquisition and the prolonged, subsequent effects on speech and language development postsurgically [38].
5. While both surgeons and orthodontists ponder the best surgical techniques and timing for producing the results everyone wants (normal craniofacial growth, normal speech and language development, happy parents, and a happy baby), some surgeons are reporting results on new surgical approaches that *may* minimize palatal scar tissue [40, 59]. Perhaps new surgical techniques will avoid the maxillary scarring caused by older techniques, e.g., the Wardill-Kilner pushback [36].

36.8 Conclusion

There is certainly reason to be optimistic that we will come closer to making reasonable decisions about the best timing for palatal surgery, taking into account *both* maxillofacial growth and speech development. More options are now available in terms of surgical techniques that may help to protect growth. Significant palatal scarring and its subsequent effects on growth are *not* inevitable consequences just because palatal surgery is carried out early enough to protect normal communication development. With all the advances in physical management of clefts, the expectation for every otherwise-normal child should be normal speech development.

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⁹ The “test” for velopharyngeal closure consisted of whether or not the speech pathologist heard a “sound difference” between open-nose and closed-nose conditions.

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37 Speech, Language, and Velopharyngeal Dysfunction: Management Throughout the Life of an Individual with Cleft Palate

JOHN E. RISKI

37.1 Introduction

One of the often-stated goals of cleft care is to establish normal speech. As providers, we attempt to achieve this goal in the face of the structural deficits of the cleft palate, a changing velopharyngeal environment, a developing dentofacial structure, and a propensity for hearing loss. We attempt to do this understanding that velopharyngeal competence is essential to normal speech learning. We are also working within a “speech readiness” period. If normal speech is not established or if maladaptive speech is not extinguished during this “readiness period,” results can be less than optimal. This chapter will review the challenges and factors that influence the outcome of managing speech, language, and velopharyngeal dysfunction throughout the life of an individual with cleft palate.

37.2 Challenges

Despite the advances in surgical technique and procedures, and the sophistication of instrumental assessment and imaging, there is little consensus on an optimal treatment protocol. A contributing factor might be the great variability in craniofacial morphology and the response to treatment in patients who have exactly the same cleft lip and palate diagnosis [1]. While we have not been able to develop a consensus on management, we are developing a clearer understanding of factors that influence our treatment. Although there is no consensus on a management program, the American Cleft Palate-Craniofacial Association has published recommended parameters of practice for the care of individuals with congenital craniofacial anomalies [2]. The consensus conference participants who developed the document espoused several fundamental principles of optimal care in-

cluding the importance of team care, parent/caregiver involvement, and longitudinal follow-up.

Hypernasality, nasal air emission, and other aberrant speech problems mark the failed primary management of the patient with cleft palate. Despite best attempts, primary palatal management is only successful in 70%–80% of individuals with cleft palate [3, 4]. That is, 20%–30% of children born with cleft palate will require secondary palate and management. Despite advances in instrumentation used to evaluate velopharyngeal function, we still have no way of predicting or identifying, at the time of initial palatal surgery, which 20%–30% of children will benefit from a pharyngoplasty. The speech pathologist must monitor emerging speech and language for any signs of nasal air escape, hypernasality, or aberrant misarticulation in order to identify children who have had a unsuccessful initial surgery, and which children are candidates for secondary surgery.

In the last 40 years, great progress has been made toward a better understanding of many aspects of the cleft lip and palate defect, but considerable study remains before there is agreement on the optimal treatment procedures. Molsted [1] provides an excellent review of the topic. Primary closure of the palate may fall into two main schools of thought. One advocates early closure of the lip and palate in order to achieve normal speech function. The other recommends delayed closure of the hard palate in order to achieve maximum growth of the maxilla. A number of inter-center and multicenter studies have been carried out recently in an effort to investigate which procedures give the best result, both esthetically and functionally. The results are ambiguous, and this has led a number of researchers to suggest that the randomized clinical trial is the only way to resolve the ambiguity. The fact that it has proved difficult to identify the optimal procedures in the field of cleft lip and palate treatment may not only be due to a less than optimal research design; a contributory factor might also be the great

variability in craniofacial morphology and in the response to treatment in patients who have exactly the same cleft lip and palate diagnosis. Intensive research has made it possible to state categorically that clefts occur due to many different factors in an interplay between genetics and environment. Therefore, it is not likely that a single gene can be responsible for clefting.

Further complicating the discussion is the topic of fetal surgery. Since scar tissue presents many problems – for instance, impairment of growth – the reduction or prevention of scar formation has long been a desirable goal. The discovery that a fetus can heal without scar formation has led to many animal experiments. The timing of the surgical intervention on fetuses is critical, since late-gestation fetuses heal with adult-like scarring. There are still many unsolved problems connected with fetal surgery, and at present prenatal surgery for repair of cleft lip and palate is not ethically defensible in humans. On the other hand, it appears that there are considerable possibilities for the reduction of human scarring after surgery with the introduction of various wound-healing medications.

Although oral clefts are among the most common birth defects, they remain a low-incidence disorder in pediatric practice. The prevalence is approximately 1:750 to 1:1,000 newborns in the Caucasian population and is even higher in other populations. It is estimated that there are 6000 new CL/P cases in the United States annually. Despite the relative prevalence of oral clefts, these patients still represent a small part of the pediatric caseload outside craniofacial centers. Hypernasality, nasal air emission, and compensatory articulation are also low-incidence speech disorders and may result from structural, neurological, or functional (learned) etiologies. The rate of *hypernasality* after initial cleft palate closure is only 20%–30% [4, 5]. Low-incidence disorders such as oral clefts and hypernasality offer few educational or clinical opportunities for developing clinical expertise. We are challenged to educate other healthcare workers about appropriate identification, diagnosis, and management.

A challenge to evaluation is that the etiologies of hypernasality and nasal airflow disorders are often occult or hidden. In recent reviews of patients receiving surgical correction for hypernasality, approximately 30% did not have a cleft palate [6, 7]. The etiology in these children is an anatomically deep nasopharynx that can be diagnosed accurately only by radiographic assessment. Normal velopharyngeal dimensions were described by Subtelny [8] and highlighted by Zemlin [9]. Further, the anatomic defect of a disproportionately deep pharynx was described by Calnan [10].

An issue in speech pathology that may have its origin in the low incidence and occult nature of the dis-

order is that noncleft hypernasality is erroneously labeled as a “voice disorder.” Labeling hypernasality a “voice disorder” is ambiguous and often has dire consequences for the child and successful management of the disorder. Labeling hypernasality as a voice disorder implies that it is a disorder of the larynx and results in delayed identification, labeling, referral, and management. Because the physical defect is not recognized, ineffective speech therapies are often poorly designed [11]. This further delays identification and referral to specialists at craniofacial teams. A cleft palate is identified by or at birth and palate closure is before one year of age [7]. In stark contrast, the average age of referral to our center for children with noncleft hypernasality resulting from Velo-Cardio-Facial Syndrome (VCFS) is 9.2 years of age! Children with hypernasality resulting from cleft palate are generally referred to the craniofacial team. In contrast, children with hypernasality with no obvious form of clefting are labeled as having a “voice disorder” and referred to an ENT, generally in a private office, who may or may not have experience evaluating or managing velopharyngeal dysfunction. This protocol followed in our school systems is a deterrent to successful identification, evaluation, and management.

Delayed management of VPI leads to increased failure of surgical intervention and refractory speech deficits. The rate of complete success when VPI is managed before 6 years of age is 90.9%. The success rate falls to 73.9% between 6 and 12 years; 70.0% between 12 and 18 years; and 47.0% after 18 years [6].

The evaluation of oral clefts and hypernasality is conducted by specialists in craniofacial clinics. However, Public Laws 94–142 and 99–457, which culminated in the Individuals with Disability Education Act (IDEA), have mandated that special services such as speech therapy be provided through specialists in schools and developmental centers. The professionals in these settings usually have limited experience with cleft-related problems because these problems usually form a very small part of their caseload. This separation of evaluation and therapy can lead to poor communication between professionals and therapy plans that do not directly address the therapy needs of the patient. There is an unquestionable need for partnerships between the evaluation centers and the settings in which the therapy is conducted.

37.3 Predicting the Need for Secondary Palate for Management

Prospective investigations designed to identify which children might benefit from primary pharyngoplasty have been unsuccessful. One investigation evaluated the speech and velopharyngeal function of 16 chil-

dren who underwent a primary Orticochea (sphincter) pharyngoplasty at the time of palate closure [12]. In the judgment of the surgeon, these children would have velopharyngeal incompetence following palatoplasty alone. Speech and velopharyngeal functioning were evaluated after five years of age. Analysis of the children's velopharyngeal function demonstrated that all had velopharyngeal competence. However, careful analysis of imaging studies demonstrated that one-half of children were demonstrating velopharyngeal closure above the point of pharyngoplasty insertion. Analysis of the children's speech demonstrated that one-fourth still had maladaptive speech problems associated with velopharyngeal incompetence. One might assume that in these children the timing of surgery was too late. In contrast, one child, whose palate was closed at 30 months, had normal speech. Closure at 30 months was obviously not too late in this child.

Mazaheri, Athanasiou, and Long [13] evaluated the usefulness of measurements taken from lateral cephalometric radiographs to predict later velopharyngeal competence. The lateral radiographs of 75 children with cleft palate were digitized for analysis. The measurements from radiographs of children who had velopharyngeal competence and acceptable speech were compared with those who presented with velopharyngeal incompetence requiring pharyngeal flap surgery or prosthesis. Soft tissue landmark points in the velopharyngeal region were digitized. Length and thickness of the soft palate and height and depth of the nasopharynx were measured. Evaluation of the growth curves of these four cephalometric variables indicated only two significant differences between children who later required pharyngeal flap surgery and those who did not. These differences were found in the growth in length of the soft palate of the cleft-palate only group and in the growth in depth of the nasopharynx of the bilateral cleft lip/palate group. Based on the present cephalometric data, it was impossible to predict at an early age those cleft lip and/or palate patients who will later require management of velopharyngeal incompetence.

Other investigators have demonstrated that a careful speech analysis has important predictive value of future velopharyngeal competence. Van Demark and his colleagues produced a number of investigations in the 1970's that valued the predictive value of the articulation test. Van Demark and Morris [14] studied the articulation test scores for 278 subjects with cleft palate and compared the results with the normative data of the Iowa Pressure Articulation Test (IPAT) and the Templin-Darley Screening test of Articulation. In addition, the predictive value of the IPAT in relation to the need for secondary management was examined. Examination of the data concerning "risk rates" indicates that the IPAT is very useful in predicting the

need for secondary management. Subjects who obtained scores of zero on the IPAT at four and one-half years of age had a 96% risk of requiring secondary palatal management.

In 1975, Van Demark, Kuehn, and Tharp [15] evaluated the articulation scores, lateral x-rays, and clinical judgments of velopharyngeal competency of 75 subjects in order to determine their predictive value. Each of the measures resulted in at least 90% appropriate predictions. However, the combination of articulation score and lateral x-ray rating appeared to be the best predictor for this particular sample in that subsequent treatment was correctly predicted for 96% of the subjects.

37.4 Predicting Future Speech Proficiency

A single type of cleft palate does not result in a homogeneous population. Clefts of the same type (e.g., unilateral cleft of the lip/palate) will have very different outcomes. Some children will develop normal speech following surgical repair of the cleft with very little other intervention. Do they have more of a chance of velopharyngeal competence, better hearing acuity, and a dental arch more conducive to normal speech? Other children develop speech and language more slowly and are ushered into early intervention programs. We still have no way of identifying which children are going to require extensive behavioral intervention without continually monitoring speech development and careful evaluation of speech and velopharyngeal function.

Although gender is not a significant variable in the outcome of surgery, it appears to be an important variable in the development of speech proficiency. Hardin and colleagues [16] examined the relative contribution of clinical data obtained during speech examinations at age 4 years and at each subsequent age through 13 years. The purpose was to evaluate the ability of data from four years of age to predict proficiency at age 14 years. All children had unilateral cleft of the lip and palate. Sixteen independent variables used in the analysis at age 4 years included four non-speech variables (gender, type and age of primary surgery, and pharyngeal flap surgery) and 12 speech measures obtained from speech examination. In subsequent analyses at ages 5 through 13 years, the rate of change between adjacent age levels for each of the 12 speech measures was also included, for 28 independent variables. The results of the investigation indicated that a large percentage of the variance in judged speech proficiency at age 14 years could be accounted for, using clinical data obtained for speech examinations at ages 4 through 13 years. Regression analyses

were performed for the total group of subjects at ages 4 through 13 years. The most efficient set of predictors accounted for 50%–75% of the variance in the dependent variable. The variable of gender was identified as the single most important predictor in nine of the 10 regression analyses, alone accounting for at least 40% of the variance in judged speech proficiency. When the regression analyses were performed separately for males and females, approximately 50%–80% of the variance was accounted for in judged speech proficiency for males and 50%–90% for females using one, two, or three of the independent variables. The most efficient set of predictors varied across age levels for both groups. In addition, these predictors differed between males and females.

37.5 Articulation Development and the Age at Palatoplasty

A goal of cleft palate habilitation is the development of normal articulation. Factors that contribute to or interfere with normal development have been studied for some time. The child's age at the initial palate closure and type of cleft have been obvious factors to evaluate.

Just as early palate closure seems to lead to better palate function, so does it appear to lead to better articulation. Hardin, Van Demark, and Morris [17] studied 48 children at 6 years of age and later in adolescence. Their results suggested the importance of establishing velopharyngeal competence by 6 years of age since resonance and articulation deficits maintained some stability after that age.

O'Gara and colleagues [18] evaluated the speech development of 23 children with one syndrome unilateral cleft of the lip and palate from 5 months to 35 months of age. The infants who had earlier palatal repair produced significantly higher percentages of oral stops (i.e., /p/, /b/, /t/) after 12 months of age than infants with later palatal repair, although there was no significant difference in mean frequency use of oral fricatives (i.e., /s/, /f/) up to 3 years of age. The results of this investigation support the earlier finding by Van Demark and Morris [14] that the use of the oral stops sounds /p/ and /b/ is a good early indicator of velopharyngeal competence.

The severity of the cleft appears to influence the number of misarticulations. Riski and De Long [5] studied the articulation of a group of children longitudinally from 3–8 years of age. They reported that the number of misarticulations on an articulation screening test increased with severity of the cleft. That is, children with bilateral cleft lip and palate had more articulation errors than children with unilateral cleft lip and palate, who had more misarticulations than

children with isolated cleft palate. As might be expected, children who also required a pharyngoplasty had more articulation errors than children who did not require a pharyngoplasty but had the same type of cleft.

The results of this investigation were supported by a later study of Finnish-speaking children [19]. Those results demonstrated that the occurrence, severity, and number of errors of all studied sounds separately or grouped increased with the severity of the cleft, being constantly greatest in the BCLP group and lowest in the CL (A) group. Further, the boys tended to have more problems than girls in producing the studied sounds correctly.

37.6 The Stability of Velopharyngeal Functioning of Time

Despite the effort to create velopharyngeal competence with primary palatoplasty, there is evidence that velopharyngeal competence is not stable and may change throughout the life of a child [20]. The roles of adenoid size and adenoid involution have been the focus of several investigations.

Gereau and Shprintzen [21] evaluated the relative size of the adenoids relative to the success or failure of primary palatoplasty. They studied velopharyngeal valving in 850 children with nonsyndromic clefts, 138 children with syndromic clefts, and a group of children without clefts. They evaluated speech relative to adenoid size. Their data suggested a strong positive correlation between the incidence of hypernasal resonance postpalatoplasty and relative adenoid size in the children with cleft palate. Velopharyngeal closure was observed consistently at the adenoids in all groups of children studied.

The importance of the adenoids in velopharyngeal closure was evaluated by Mason and Warren [22]. Specifically they evaluated the phenomenon of gradual development of hypernasality as a result of adenoidal involution. They reported that hypernasality developed in two patients from a sample of 122 with repaired cleft palate. They identified three patterns of velopharyngeal closure against the adenoid mass from lateral radiographs.

One small-sample-size study does not support the contribution of adenoid involution to developing velopharyngeal incompetence in children with cleft palate. Van Demark, Hardin, and Morris [23] compared longitudinally perceptual ratings of articulation defectiveness, nasality, and velopharyngeal competency in 13 subjects who required secondary palatal management after age 10 with a second group who did not require a pharyngoplasty. Perceptual data when examined longitudinally did not adequately discriminate between subjects who at one time achieved

velopharyngeal closure but who ultimately required secondary management and those patients who needed no further treatment. A decrease in articulation scores and an increase in severity of nasality and articulation defectiveness over time indicate that patients are at risk for secondary management. Evaluation of lateral x-rays indicated that (1) those in the group who required secondary operations demonstrated more variability in velopharyngeal closure than those in the comparison group, who required no secondary operations, and (2) adenoidal involution did not appear to be a significant factor.

Morris, Wroblewski, Brown, and Van Demark [24] evaluated the role of hypertrophied adenoids and any role they might have in maintaining velopharyngeal competence. Thirty-nine subjects were selected from a large longitudinal study based on availability of lateral still x-ray films taken in series from 5 to 16 years of age. Ratings of velar-pharyngeal contact and ratings of adenoid size were obtained from the films. The obtained data indicated the expected decrease in adenoid size. All patients maintained velopharyngeal competence during the period of study. However, three of the 39 subjects were judged to show loss of such contact during the period of study, and an additional four had surgery for velopharyngeal incompetence after the completion of the study. All seven appeared to show significant deterioration of velopharyngeal status in middle or late adolescence.

In a larger study of 121 children born with cleft (lip) palate, Riski, Stewart, Mason, and Simms [25] evaluated velopharyngeal competence relative to adenoid involution. They reported that 11/121 (9.1%) developed velopharyngeal incompetence with adenoid involution. Velopharyngeal function was evaluated with clinical and instrumental tools. Each of the 121 children initially demonstrated velopharyngeal closure characterized by the absence of any nasal air escape or hypernasality. However, at an average age of 5.7 years nasal air escape was first detected. Lateral radiographs were taken at that time to monitor adenoid size. At an average age of 8.6 years, the adenoids had involuted 4.64 mm, leading to significant velopharyngeal incompetence requiring management.

The importance of the adenoid mass for velopharyngeal competence in children with cleft palate was also highlighted in a longitudinal study reported by Haapanen, Veija, and Pettay [26]. They evaluated two groups of children with cleft palate. One group of 24 children had an adenoidectomy at the time of palatoplasty, and a second group of 25 children with cleft palate did not have an adenoidectomy. They found that hypernasality and nasal air escape occurred significantly more frequently in the group of children who had an adenoidectomy than in the group of children who did not. The groups did not differ when it

came to articulation errors associated with velopharyngeal insufficiency.

37.7 Age at Pharyngoplasty

Given that 20% of children will demonstrate velopharyngeal incompetence following palatoplasty, considerable effort has been expended to identify the ideal age for secondary cleft palate management. Many authors of studies of pharyngoplasties have commented on the most appropriate age for intervention. Most have reported better results in younger patients. An early study by Moll, Huffman, Lierle, and Smith [27] reported better results in patients younger than 15 years of age. Leanderson et al. [28] evaluated the results of secondary management by pharyngeal flap in 124 patients with cleft palate. They reported the best results between 5 and 6 years of age. They observed continued speech improvements for several years after surgery in young patients. No improvement was found in older (over 25 years) patients.

Riski [4] reported on longitudinal data of speech sound acquisition and oral-nasal balance in 52 children who had had pharyngeal flaps for velopharyngeal incompetency. These data were compared to similar data for a group of 48 children with cleft palates who had not required secondary management. Comparisons were made also between the pre- and post-surgical performance of the group who had had pharyngeal flaps. Further comparisons were made between children who had secondary surgery before 6 years and after 6 years of age. Development of acceptable resonance and articulation patterns was slower in children who required pharyngeal flaps compared to those who did not. Further analysis showed that the post-pharyngeal flap group demonstrated acceleration in the acquisition of acceptable sound production in the year immediately following surgery. The data suggested that children who had flaps before 6 years of age made faster gains in the development of articulation and acceptable resonance than did children who were treated after the age of 6 years.

Van Demark and Hammerquist [29] observed a trend for poorer articulation when a pharyngeal flap was done after 10 years of age. Surprisingly, when studied at 10 years of age, children with pharyngeal flaps done earlier than 5 years of age had poorer velopharyngeal competence ratings than children with pharyngeal flaps done later than 5 years of age. They concluded that the optimal time for pharyngeal flap was between 4 and 10 years of age.

Seyfer, Prohazka, and Leahy [30] reported the results of superiorly based pharyngeal flaps in 39 patients. All 11 (100%) patients under 6 years of age im-

proved. The average improvement was 1.1 on a 3-point scale. In contrast, 74% of the 28 patients over 6 years of age improved by 1.4 on the same 3-point scale. The difference was not statistically significant, however; in addition, no significant difference was observed between the improvement for children less than 2 years old (including primary pharyngeal flaps) and those over 2 years old.

Van Demark and Hardin [31] evaluated 129 patients with pharyngeal flaps. Articulation improved 15% postoperatively and was only 90% correct by 16 years of age. That is, speech was not normal in the oldest group studied. Although articulation ratings were better for patients receiving flaps earlier than 4 years of age or between 4 and 5 years of age, there was no difference in nasality when patients receiving flaps early and late were compared. They concluded that age at which pharyngeal flap surgery was performed was not critical in speech outcome.

A large series of children who had a sphincter pharyngoplasty was studied by Riski et al. [6] using aerodynamic and imaging data. In this series, 109 of 139 patients demonstrated resolution of VPI. Sixteen of the initial failures were revised. The revision was successful in eight of 16, yielding an overall success rate of 117 of 139 (84.2%).

Additional analysis of the series demonstrated that patients with mild hypernasality (90.38%) were treated more successfully than those with severe hypernasality (71.26%). Preoperative velopharyngeal orifice area, estimated by aerodynamic testing, influenced surgical outcome to some degree. The 64 patients with normal postoperative resonance demonstrated a preoperative gap of 9.9 mm²; the 11 hypernasal patients had a larger preoperative gap of 12.2 mm², and the four hyponasal patients had a much smaller preoperative gap of 5.0 mm².

Age was a more significant determinant of outcome than was size of velopharyngeal gap. Sixty children under age 12 had a success rate of 84% and a preoperative area of 9.6 mm². Twenty patients over 12 years had a success rate of only 55% and a similar preoperative orifice area of 10.9 mm². A closer analysis of the age information revealed that the rate of complete success when VPI is managed before 6 years of age is 90.9%. The success rate falls to 73.9% between 6 and 12 years, 70.0% between 12 and 18 years, and to 47.0% after 18 years.

Finally Losken et al. [32] found no statistical difference in the average age at initial sphincter pharyngoplasty in patients who required a revision (6.8 years + 4.48) compared to those who did not require revision (7.4 years + 4.4).

37.8 The Adult Patient

Although VPI is usually diagnosed and treated in childhood, it is not uncommon for adults to present for management. Younger and Dickson [33] treated eight adults with residual VPI following cleft palate repair as children. A superiorly based pharyngeal flap was used in each case. They reported "significant subjective and objective improvement," although they reported no data.

A more controlled study was reported by Hall, Golding-Kushner, Argamaso, and Strauch [34]. Twenty adult patients received a superiorly based pharyngeal flap. The authors reported normal resonance in 15 patients, hyponasality in three, and residual hypernasality in two. The heterogeneity of the sample made conclusions difficult. Of the 20 patients, 13 had unrepaired clefts, two had speech bulbs, and one had a failed pharyngeal flap.

37.9 Primary Versus Secondary Pharyngoplasty

Several studies have been reported of pharyngoplasty used during primary repair of the palate. The pharyngeal flap is advocated to provide additional tissue when the palatal cleft is extremely wide [35]. Both the pharyngeal flap [36] and the sphincter pharyngoplasty [12] have been used at the time of primary palatal closure in an attempt to increase velopharyngeal competence. Both procedures have a higher rate of success than a palatoplasty alone. An inferiorly based flap combined with a palatoplasty has been shown to eliminate hypernasality in 94% of patients [36]. A sphincter pharyngoplasty combined with a palatoplasty has been shown to have a 100% success rate [12]. Controversy still exists about using any secondary procedure with primary palatoplasty, because only 20% to 30% of patients will eventually require pharyngoplasty and because of the potential obstruction. The ability to predict which infants will eventually require a pharyngoplasty appears to be poor. One surgeon attempted to identify infants who would eventually need a pharyngoplasty by visual inspection of the nasopharynx at the time of palatoplasty. Sixteen of these children were later evaluated. The pharyngoplasty appeared necessary in only 50% [12]. The remaining children achieved closure above the pharyngoplasty at the adenoids.

37.10 Speech Therapy

Our ability to evaluate velopharyngeal function is much more exact than our ability to treat velopharyngeal dysfunction and associated misarticulations with speech therapy. Many procedures have been proposed for treating velopharyngeal dysfunction, but there is little evidence that any of these techniques actually improves velopharyngeal function [37]. Currently, there seems to be a proliferation of oral motor exercises without any evidence supporting these procedures. Evidence from the literature suggests that nonspeech, oral motor exercises have little basis in fact. Oral motor exercises isolate oral movements by manipulating the oral structures either manually or with instruments such as toothbrushes. In contrast, speech is a coordinated process incorporating coordinated movements of respiration, laryngeal, velopharyngeal, and oral articulatory gestures. Proponents of oral motor exercises often proposed that nonspeech oral movements must develop before learning speech oral movements. Research suggests however that they develop in parallel.

Moore and colleagues have published a number of investigations evaluating whether speech emerges from earlier-appearing oral motor behaviors. Moore and Ruark [65] designed an investigation to quantify the coordinative organization of mandibular muscles in toddlers during speech and nonspeech behaviors. Seven 15-month-olds were observed during spontaneous production of chewing, sucking, babbling, and speech. They found that mandibular coordination across these behaviors was quite different from that observed for other behaviors, even in the earliest stages of true word production. Production of true words was predominantly characterized by relatively stronger coupling among all mandibular muscles compared to earlier emerging chewing and sucking. Variegated babbling exhibited stronger coupling than reduplicated babbling, as well as chewing and sucking. They concluded that their findings did not support the suggestion that speech coordination emerges from earlier-appearing oral motor behaviors. Further, they concluded that the finding of coupled activation among mandibular antagonists during speech paralleled earlier comparisons of adult speech and nonspeech behaviors.

In a follow-up investigation, Ruark and Moore [38] reported on an investigation that was designed to quantify the coordinative organization of lip muscle activity of 2-year-old children during speech and nonspeech behaviors. They recorded electromyographic (EMG) activity from the right upper and lower lip activity of seven 2-year-old children during productions of chewing, syllable repetition, lip protrusion, and speech (repeated two-word utterances)

tasks. Task comparisons revealed that the coordinative organization of upper and lower lip activity is task-specific; different coordinative strategies are employed for different tasks. They concluded that the finding of coordinative elements of the perioral system of 2-year-olds are task-specific, and extends the results of previous studies of adults and children, where task-specific coordinative strategies were employed by the mandibular and perioral. Further, they concluded that the task-dependent coordination of the perioral system of 2-year-olds supports the notion that developing speech and earlier-developing oromotor behaviors (i.e., sucking, chewing) are mediated by different control mechanisms.

Parallel to the findings of parallel mechanisms for speech and nonspeech production is the report from Benson et al. [39], who found a parallel rather than serial model of auditory speech and nonspeech perception. They identified candidate brain regions constituting a neural network for preattentive phonetic perception with MRI and multivariate multiple regression of imaging data. They used stimuli that were contrasted along speech/nonspeech, acoustic, or phonetic complexity (three levels each) and natural/synthetic dimensions. They found that seven distributed brain regions' activity correlated with speech and speech complexity dimensions, including five left-sided foci [posterior superior temporal gyrus (STG), angular gyrus, ventral occipitotemporal cortex, inferior/posterior supramarginal gyrus, and middle frontal gyrus (MFG)] and two right-sided foci (posterior STG and anterior insula). In contrast, only the left MFG discriminated natural and synthetic speech. They concluded that the data supported a parallel rather than serial model of auditory speech and nonspeech perception.

A place for isolated oral motor exercises has yet to be defined. There is the potential that maladaptive speech habits will continue and become more ingrained as ineffective and unproven exercises continue.

Continuous positive airway pressure (CPAP) therapy can be used to reduce hypernasality by elevating the air pressure in the nasal cavities during speech [66]. The purpose of this study was to determine whether increased intranasal air pressure loads the major muscle of velopharyngeal closure, the levator veli palatini. Nine subjects, four with cleft palate and five without cleft palate, were studied. Electromyographic activity was measured from the levator veli palatini muscle with several levels of air pressure delivered to the nasal cavities using a commercially available CPAP instrument. It was found that levator veli palatini activity was significantly greater for the positive air pressure conditions than for the atmospheric pressure conditions for both subject groups. This indicates

that the levator veli palatini muscle acts against the resistive load produced by the increased intranasal air pressure. The results support the use of CPAP therapy as a method of resistance exercise for strengthening velopharyngeal closure muscles.

Very few studies have evaluated speech therapy treatment strategies for a child with cleft palate. Pamplona, Ysunza, and Espinosa [40] compared the amount of speech therapy time required to correct compensatory articulation for two types of speech therapy. One group was treated with a phonologic-based intervention, and the second modality was treated with an articulatory or phonetic approach.

Twenty-nine patients ranging from 3 to 7 years were treated. Fifteen patients were included in the first group (control) and received articulatory therapy, and 14 patients were included in the second group (active) and received phonologic therapy. The median age of each group was similar; that of the control group was 54 months old, and that of the active group was 55.50 months old. The speech pathologist in charge of the speech therapy was the same in all cases. A blind procedure was utilized whereby each patient was evaluated independently by two speech pathologists every three months until both examiners were convinced that compensatory articulation had been completely corrected.

The mean total time of speech therapy required for the normalization of speech in the two groups of patients was compared. The mean total time of therapy was 30.07 months in the control group and 14.50 months in the active group. A Student's *t*-test demonstrated that the total time of therapy was significantly reduced ($P < 0.001$) when a phonological intervention was utilized.

37.11 Role of Parents in Speech Therapy

The child developing speech and language might have a parent who is actively involved in providing speech and language stimulation, or one who is passive. The evidence from studies of speech and language intervention is clear that the child with active parent participation develops better language skills. Studies of children with cleft lip/palate as well as other disabilities demonstrate that it is paramount that clinicians incorporate the parent as an active participant in speech therapy and speech and language stimulation.

Pamplona and Ysunza [41] and Pamplona, Ysunza, and Uriostegui [42] investigated whether including the mother as an active participant in speech therapy sessions would improve the language development of children with cleft palate who also had additional language delays. One group was treated by the speech pathologist alone (control group), and a second group

was treated by the speech pathologist but was also accompanied by their mothers (experimental group). Both groups were evaluated before and after treatment to evaluate the advance of each group. The patients accompanied by their mothers had significantly better language skills compared with patients treated without their mothers. They concluded that the results supported the statement that language development is related to mother-child mode of daily life interaction in children with cleft palate.

Additional support and evidence for parent involvement comes from Broen, Doyle, and Bacon [43]. They described the changes in a 3-year-old child's production of speech during a period of diagnostic therapy, and the changes that occurred following the fitting of a speech prosthesis. The child's mother served as the primary intervener, guided by a speech-language pathologist. The mother was able to change the child's speech so that more of her productions were at a correct place of articulation. After structural management, nasal and glottalized productions disappeared from the child's speech, but glottal stops did not.

Support for the importance of parent participation is also found in investigations of children with other disorders. Janjua, Woll, and Kyle [44] evaluated language development in severe and profound hearing-impaired children and the type of parental interactions. They found that children who were directly involved in an activity with their parents develop better language, as were children who performed the task after their parents demonstrated it to them. The authors recommended that parents encourage child participation and use more contingent and child-centered interaction. In addition, they found no significant differences between oral and bilingual families in terms of quality of interaction.

Girolametto et al. [45] studied 10 English-speaking mother-child dyads and 10 Italian-speaking mother-child dyads. All 20 children were late talkers who showed delays in expressive vocabulary development but age-appropriate cognitive and receptive language skills. The authors found some cultural differences in language learning and concluded that their results support the use of language interventions based on increasing maternal responsiveness for these children at the one-word stage of language development.

37.12 Language Development and Learning

The school-aged child with a cleft palate presents the clinician with unique considerations beyond articulation and velopharyngeal function. Evidence first surfaced in the 1970's that children with cleft palate

might have educational problems that separate them from children without clefts. Richman [46] evaluated 44 cleft lip and palate/palate only children who were individually matched with 44 normal school children on the basis of sex, age, grade, intelligence, and socioeconomic status. All children received behavioral ratings by classroom teachers, and achievement test scores were obtained. The children with cleft palate were rated by teachers as displaying significantly greater inhibition of impulse (internalizing behavior). Further, the children with clefts also scored significantly lower on overall basic skills achievement tests. At that time, Richman suggested that cleft children might be responding to the social-behavioral environment, which may include negative social responses from others.

Later, Richman [47] examined patterns of cognitive ability in 57 cleft lip and palate children with verbal deficits, but without general intellectual impairment. He was attempting to answer the question of whether the verbal disability displayed by these children was related primarily to a specific verbal expression deficit or a more general symbolic mediation problem. Two groups of children were identified based on performance on cognitive tasks which required verbal mediation strategies without requiring vocal responses. The children with only a verbal expression problem performed significantly better on tasks requiring categorization and associative reasoning, although there were few apparent differences on memory items. Those children with a verbal expression deficit displayed both an underlying symbolic mediation deficiency and learning disabilities. There was a higher proportion of cleft-palate only children in the more severe group.

Richman and Eliason [48] reviewed the research on intelligence, achievement, behavior, and personality of children with cleft lip and palate. Studies of intellectual functioning demonstrated that the general intelligence of cleft populations is relatively normally distributed with group mean IQ scores within the average range. There is some suggestion of a higher frequency of depressed verbal intellectual functioning relative to visual-motor intelligence. Factors that appear to affect IQ levels are presence of other congenital anomalies, speech and hearing deficiency, and low-incidence cleft type by sex occurrences. There is evidence that a high percentage of cleft children are underachievers. Personality and behavioral studies do not suggest significant psychopathology, although there is evidence of behavioral inhibition, concern regarding appearance, and decreased expectations by teachers and parents.

Richman and his colleagues first delineated specific reading and learning disabilities in children with isolated clefts. Richman and Eliason [48] studied children

with cleft lip and palate and cleft palate only. Both groups presented with reading disability and were matched for intelligence, age, sex, and reading level. The two groups were compared on reading and neuropsychological test variables. Subjects included 14 males and 10 females of each cleft type ranging in age from 8 to 13 years. The results showed a significant difference between groups on most language measures and differences in reading comprehension and type of reading errors. These results suggested that children with cleft palate-only constitute a language-disorder group with more severe reading disabilities. Children with cleft lip and palate are more likely to have verbal expressive deficits and milder reading problems, possibly related to peripheral speech mechanisms.

Richman et al. [49] were then able to delineate the incidence of reading disabilities in children with isolated cleft palate. They examined 172 elementary school children with cleft lip and palate (CLP) or cleft palate only (CPO). Approximately 35% of the sample displayed a moderate degree of reading disability, and 17% of the group exhibited severe reading disability. Reading disability was more prevalent at younger ages, presumably because of peripheral speech deficits. For older children, those with CLP displayed an incidence of reading disability similar to the general population (9%). However, children with CPO exhibited a much higher rate of reading disability (33%). There was no difference between genders in the prevalence of reading disability in this sample. This study supported their earlier research that suggested children with CPO may be more likely to experience general language disorders leading to long-term reading disabilities. Children with CLP appear to manifest reading problems that tend to resolve with increased age.

Investigations that are more recent have highlighted the elevated incidence of learning disabilities in children with clefts, especially those with isolated cleft palate. Broder et al. [50] examined the prevalence of learning disability (LD), level of school achievement, and prevalence of grade retention by type of cleft and gender at two craniofacial centers. They studied 84 consecutively evaluated patients from one center who were matched by cleft type, age, and gender with 84 patients evaluated at a second center. They found that 46% of subjects with cleft had LD, 47% had deficient educational progress, and 27% had repeated a grade (excluding kindergarten) in school. Males with cleft palate only (CPO) had a significantly higher rate of LD than any other subject group. Males with CPO and females with CLP were more likely to repeat a grade in school than were females with CPO and males with CLP. They concluded that children with cleft are at risk for learning disability, low school achievement, and grade retention.

These investigations have prompted a series of studies looking at preschool children. Scherer and D'Antonio [51] used a parent questionnaire as a component for screening early language development of children 16 to 30 months of age with cleft lip and palate. Thirty nonsyndromic children with cleft lip and palate and 30 children without clefts received the MacArthur Communicative Development Inventory: Toddler (CDI: Toddler), administered by a pediatrician. In addition, a speech-language screening was performed by a speech-language pathologist. Results of the two assessments indicated that the CDI: Toddler was a valid screener of language development compared to a comprehensive speech-language screening. They reported differences between the cleft and noncleft groups and found evidence of delays in expressive language development in the children with cleft lip and palate.

The source of language-learning disabilities was investigated by Ceponiene et al. [52].

They investigated persistence of auditory short-term memory (STM) that is implicitly involved in language-specific perception in children with clefts, grouped using fine-graded cleft classification. Cortical evoked potentials were recorded in 78 children with nonsyndromic oral clefts and in 32 healthy peers. A mismatch negativity (MMN) potential that indexes preattentive detection of change in auditory input was obtained in response to tone sounds. In order to test durability of short-term memory traces, sounds were presented with three stimulation rates. With slowest stimulation, MMN amplitudes were reduced in cleft children compared to the healthy peers ($P < 0.00065$). Only cleft-lip children did not significantly differ from controls. Among isolated palatal clefts, the more posteriorly delimited the cleft was, the smaller was the amplitude of MMN. MMNs of smallest amplitudes were obtained in the subgroup of complete unilateral cleft of lip and palate. The authors concluded that the reduced MMN amplitudes found in cleft children imply deficiency in auditory STM trace maintenance. This dysfunction is likely to contribute to their language and learning disabilities. The MMN diminution with shorter/more posterior clefts suggests that differences in auditory cortex function are one of the underlying mechanisms of the cleft type-malocclusion association.

In a follow-up study, Ceponiene et al. [53] investigated preattentive auditory discrimination, reporting that it plays a significant role in language acquisition and usage. They evaluated infants with different cleft types. A mismatch negativity (MMN) component of brain evoked potentials, which indexes preconscious sound discrimination, and brain responses to rare sine-wave tones were recorded in 12 healthy infants and 32 infants with oral clefts at the ages of 0 and

6 months. Infants with clefts were subdivided into two categories: those with cleft lip and palate (CLP) ($n=11$ at birth, $n=6$ at the age of 6 months) and those with cleft palate only (CPO) ($n=17$ at birth, $n=8$ at the age of 6 months). At both ages, brain responses to rare sounds tended to be smaller in both cleft subgroups than in healthy peers. However, in the latency range of 300 to 500 ms, the MMN was significantly smaller in infants with CPO. In infants with CLP, the MMN was comparable to that of healthy infants. Differences in auditory discrimination between infants with CLP and CPO, as reflected by MMN, were detectable at birth and persisted into later infancy. This pattern parallels known behavioral differences between children with these cleft types. The authors concluded that brain responses to rare sounds, in contrast, had no differentiative power with respect to the cleft type.

37.13 Preschool and School-Aged Investigations

Several investigators have evaluated language functions in infants, preschool and school-aged children. Neiman and Savage [54] obtained reports from caregivers to describe the developmental status of infants and toddlers with clefts. They studied 186 infants and toddlers with cleft lip, cleft palate, and cleft lip/palate, using the Kent Infant Developmental Scale and the Minnesota Child Development Inventory, both caregiver reports. Reports were obtained at one of the following age categories: 5 months, 13 months, 25 months, and 36 months. Data were analyzed in separate two-between ANOVAs (age $\times$ cleft type) for each developmental domain according to developmental assessment measure. Further, results were examined relative to the normative sample.

Lower-motor and self-help developmental quotients were lower at 5 months compared to the 13-month-old level. The 5-month-old infants with cleft exhibited 'at-risk/delayed' development on the motor, self-help, and cognitive domains compared to the normative sample. Infants at 13 and 25 months were within normal limits in all developmental domains, with the exception of the 13-month-old infants with cleft palate, who demonstrate 'at-risk' development in the motor domain. Toddlers with cleft palate exhibit 'at-risk/delayed' development in the expressive language domain at 36 months.

Early delays were also reported by Broen et al. [43]. They studied the early cognitive and linguistic development of 29 children with cleft palate compared to that of 29 children without clefts. Measures included the Mental scale of the Bayley Scales of Infant Development, the Minnesota Child Development Inventory, Mean Length of Utterance, and words acquired by

24 months. Children with cleft palate, although well within the normal range, performed significantly below the children in the noncleft group on the Mental Scale of the Bayley Scales of Infant Development, some subscales of the Minnesota Child Development Inventory, and words acquired by 24 months. Differences observed in the cognitive development of children with and without cleft palate were verbal as opposed to nonverbal (i.e., linguistic in nature) and were related to hearing status at 12 months and velopharyngeal adequacy.

Chapman et al. [55] examined the conversational skills of preschool and school age children with cleft lip and palate during interactions with unfamiliar adults. Standardized measures of speech and language were administered, and ratings of resonance were obtained. Comparisons were made between the children with cleft lip and palate and their same-age peers on measures of conversational participation and a standardized test of pragmatic skills. Twenty children with unilateral cleft lip and palate (10 preschoolers and 10 school-age children) and 20 noncleft peers were matched for gender, age, and socioeconomic status.

Paired t-tests revealed no significant differences between the preschool and school-age children with cleft lip and palate and their noncleft peers in level of conversational participation. However, individual child comparisons revealed less assertive profiles of conversational participation for 50% of the preschool and 20% of the school-age children with cleft lip and palate. The investigators concluded that children with cleft lip and palate may show a less assertive style of conversational participation, at least during the preschool years. Therefore, craniofacial team evaluations should include examination of conversational competency, particularly for children who are demonstrating difficulty with other aspects of speech, language, or social development.

The work of Speltz and colleagues suggest that the differences between cleft lip/palate and cleft palate-only children may not be revealed until preschool or school age. Speltz et al. [56] used cognitive and psychomotor tasks to evaluate 29 infants with cleft lip and palate (CLP), 28 infants with cleft palate only (CPO), and a demographically matched comparison (COMP) group of 69 infants. The children were studied at ages 3, 12, and 24 months. The purpose was to examine predictors of cognitive status at age 24 months in the cleft group. Infants were administered the Bayley Scales of Infant Development (BSID), mother-infant interactions were observed, and medical records were reviewed. The CLP and CPO groups scored lower than the COMP group on the BSID, but did not differ from one another. Cleft group infants scored lower than COMP group infants on BSID items assessing nonver-

bal and expressive language skills. Quality of maternal interaction predicted the 2-year Mental Development Index (MDI) scores of infants with clefts. They concluded that infants with clefts show relative deficits in cognitive and psychomotor development and that cognitive deficits are apparent in nonverbal as well as verbal areas of performance.

37.14 Velo-Cardio-Facial Syndrome

Velo-Cardio-Facial Syndrome (VCFS) was first described by Shprintzen et al. [57] in 1978. They described a pattern of similarities among 12 patients including overt or submucous clefts of the secondary palate, ventricular septal defects, typical facies, and learning disabilities. Since that time we have learned that both DiGeorge Syndrome and VCFS have a microscopic deletion of chromosome 22q11.2. Treating the speech, language, and velopharyngeal dysfunction in these patients present us with special challenges beyond those of patients with clefts and are considered separately.

Golding-Kushner, Weller, and Shprintzen [58] first described a rather distinctive pattern of language disorders and personality characteristics when they described language, academic, and psychological profiles of 26 patients with velo-cardio-facial syndrome. The characteristics continued through the course of development from initial language acquisition through childhood and adolescence.

Gerdes et al. [59] suggested that the global delays and the variations in intelligence found are directly associated with the 22q11.2 deletion and are not explained by physical anomalies such as palatal defects or cardiac defects, or therapeutic interventions such as cardiac surgery. They reported that all of the children had late onset of verbal speech and behavioral disorders, including disinhibition and attention disorders. They strongly encourage early intervention services beginning in infancy to address the delays in gross motor skills, speech and language, and global developmental delays.

The observations of Gerdes et al. [59] were supported by Scherer et al. [60]. They described the speech and language development of four children with VCFS studied longitudinally from 6 to 30 months of age and compared their performance with three groups of children: (1) normally developing children, (2) children with cleft lip and palate, and (3) children with isolated cleft palate. The data demonstrated that young children with VCFS show a receptive-expressive language impairment from the onset of language. Further, speech and expressive language development were severely delayed beyond a level predicted by their other developmental or receptive language per-

formance. The children with VCFS showed severe limitations in speech sound inventories and early vocabulary development that far exceeded those shown by the children with cleft lip and palate and children with isolated cleft palate. The authors concluded that young children with VCFS emerge from a critical speech and language-learning period with severe limitations in their communicative abilities. They recommended additional studies to describe the later course of these early speech and language impairments and to explore the relationship to learning disabilities described for older children with VCFS.

More recently Murphy et al. [61] investigated the presence of 22q11.2 deletion in individuals with learning disabilities. They evaluated 265 people with learning disability residing in two learning disability hospitals in South Wales. Individuals were selected for inclusion if they fulfilled any of the following criteria: psychotic disorder (schizophrenia or affective disorder), family history of psychotic disorder, cleft palate and/or lip, congenital heart disease, broadly defined facial dysmorphism, or a history of hypocalcaemia. Fluorescence in situ hybridization studies were performed on 74 selected individuals. Cytogenetic analysis revealed that two individuals demonstrated a previously undetected chromosome 22qII deletion. A third person demonstrated a previously undetected cytogenetically visible deletion on chromosome 15. The authors concluded that VCFS appears to be etiologically significant in a proportion of individuals with idiopathic learning disability, especially in those where psychosis is associated with mild learning disability. They suggested that clinicians should consider a chromosome 22qII deletion in people who meet selection criteria.

Citing the paucity of investigations evaluating education interventions, Kok and Soman [62] reported their investigation of interactive computer-based instruction on the development of reading, language, spelling, and number skills. They recorded any positive effects of computer-based instruction on students' self-esteem, motivation, and competence in computer operational skills. They reported that the students were enthusiastic about the system, developed an interest in reading, and transferred remedial instruction to classroom performance. Comparison of pre- and post-test results indicated significant improvement in reading ability as measured using Neale Analysis.

Later, Scherer, D'Antonio, and Rodgers [63] described communication profiles in children with VCFS compared with children with Down syndrome. Four children with VCFS and four children with Down syndrome underwent cognitive and speech and language assessment. Communication profiles of children with Down syndrome showed a flat profile,

indicating all measures were similar and delayed relative to chronological age. Children with VCFS showed vocabulary, pattern of sound types, and Mean Babbling Length below cognitive and other language ages. The authors concluded that communication profiles of children with VCFS differed qualitatively and quantitatively from those of children with Down syndrome and supported the hypothesis that some children with VCFS present with a profile of communication impairment that may be distinctive to the syndrome.

The source of the learning difficulties and speech and language was investigated by Cheour et al. [64]. They evaluated the duration of auditory sensory memory, which is of central importance to speech perception and understanding. They used mismatch negativity (MMN), an attention independent event-related potential which provides an objective electrical index of auditory sensory memory. They reported that the duration of this memory span is considerably shorter in 6–10-year-old children with VCFS than in healthy controls. They concluded that the language-related problems encountered in children suffering from VCFS are likely to be caused also by central nervous system (CNS) dysfunctions.

37.15 Summary

Individuals with cleft palate face many obstacles to normal speech and language throughout their life. One of the first challenges is to identify knowledgeable and skilled caregivers. Although clefting is a frequent birth defect, it is a low-incidence disorder among pediatric disorders. This can limit the number of training programs and thus the number of treatment centers with skilled caregivers. Further, noncleft hypernasality is labeled a "voice disorder". This delays efforts to refer children with noncleft hypernasality to craniofacial teams for appropriate assessment and management. Considerable effort is needed to educate caregivers (especially speech-language pathologists, otolaryngologists, and pediatricians) that these types of problems are best evaluated and managed by craniofacial teams.

As clinicians, we are unable to predict, at the time of palatoplasty, those infants who will need secondary palatal management. Further, velopharyngeal function can be unstable in some children. As the child grows, the nasopharyngeal dimensions can become compromised as craniofacial structures grow and adenoids involute. This can lead to compromise of velopharyngeal function over the life of a child. Careful monitoring of emerging speech and velopharyngeal function remains paramount to early identification of VPI and appropriate management. Earlier

identification and management results in more successful outcomes.

The development of a single standard of treatment has been elusive. Speech-language pathologists are challenged to institute therapy regimens that are evidenced-based. There does not seem to be any evidence supporting oral motor exercises that isolate oral movements from other vocal tract structures used in speech. However, velopharyngeal exercise using CPAP and speech therapy with a phonologic approach hold some promise.

Finally, infants and children with clefts are known to have language and learning difficulties. Evaluation of emerging language in infants and language/learning skills in school-age children is indispensable. Programs that include a strong parental involvement seem to demonstrate more success than programs that do not.

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38 Prosthetic Speech Appliances for Patients with Cleft Palate

MOHAMMED MAZAHERI

Cleft lip and palate habilitation began as an individual enterprise. The development of knowledge of the nature of the defect and of the remedial measures that were helpful in allowing these patients to achieve more normal participation in community life attracted additional specialists to aid in their habilitation. The need for communication and understanding among these interested disciplines has stimulated a rapid growth of cleft palate teams during the past 15 years. The need for this concerted effort is real. The specialists comprising the cleft palate teams differ in their training and experience, which has led to the development of different treatment philosophies.

In our group, the surgeon plays a dominant role [1], because we believe that the reconstruction of a cleft palate is primarily a surgical challenge and an area where competent surgeons are capable of offering even more services to patients with cleft lip and palate than are universally practiced. Along with this philosophy, we maintain that a group enterprise by professionals is necessary in a complete habilitation program. In many areas of the world, trained personnel are not available to assist the cleft palate patient in all of his or her needs, and the surgeon has been forced to devise ingenious and occasionally extensive surgical procedures.

Cleft palate surgery is not a stereotyped exercise, but rather a service demanding an assessment of all the factors presented by each patient and a reparative surgical plan based on proven principles. The majority of cleft palates can be reconstructed by trained surgeons to enable the patient to develop acceptable speech.

Many clefts of the hard palate can be closed by a vomer flap [5–25], and clefts of the soft palate can be closed by median suture with a good anatomical and functional result. The wide cleft and the short palate demand additional attention. Additional length may be gained by the Dorrance or a V-Y type retropositioning operation. The raw nasal surface may be cov-

ered with a skin graft [3] or nasal mucosa [2], or an island flap of palatal mucosa [22, 23]. The incompetent palato-pharyngeal valve can be augmented by a pharyngeal flap, either as a primary [24] or secondary procedure. The need for additional tissue in a wide cleft can be satisfied by single or double regional flaps.

Associated with these surgical advantages now available to the cleft palate patient, there has also been a need for cleft palate prostheses. The trained prosthodontist has methods at his command to assist both the surgeon and the patient. Mutual understanding and restraint develop among the specialists in a well-organized team, to the benefit of the patient.

Since the early 16th century, dentists have been making appliances designed to close defects in the hard palate (Fig. 38.1). Like their predecessors, prosthodontists today are also vitally interested in helping the individual with an oral cleft. The prosthetic speech appliances they make are recommended as a preliminary or secondary treatment for many patients with cleft palate. The terms “obturator” and “speech appliance” will both be used in this chapter. They are frequently used synonymously, but it has become the custom to apply the term “obturator” to a device used in treating acquired defects, and “speech appliance” to one employed in treating congenital clefts.

The design and construction of speech appliances and obturators have changed much in the last 20 years, mainly due to improved materials and methods. We have, for example, better wax, acrylics, and impression materials and superior stone and plaster for the investment process. We also have benefited from improved diagnostic tools for evaluating the results of prosthetic treatment. Greater coordination of interdisciplinary team efforts has helped establish a more ideal prosthetic concept, one that ensures that fixed or removable partial denture prostheses are managed so as to preserve the integrity of all remaining teeth and the surrounding soft and hard oropharyngeal structures.

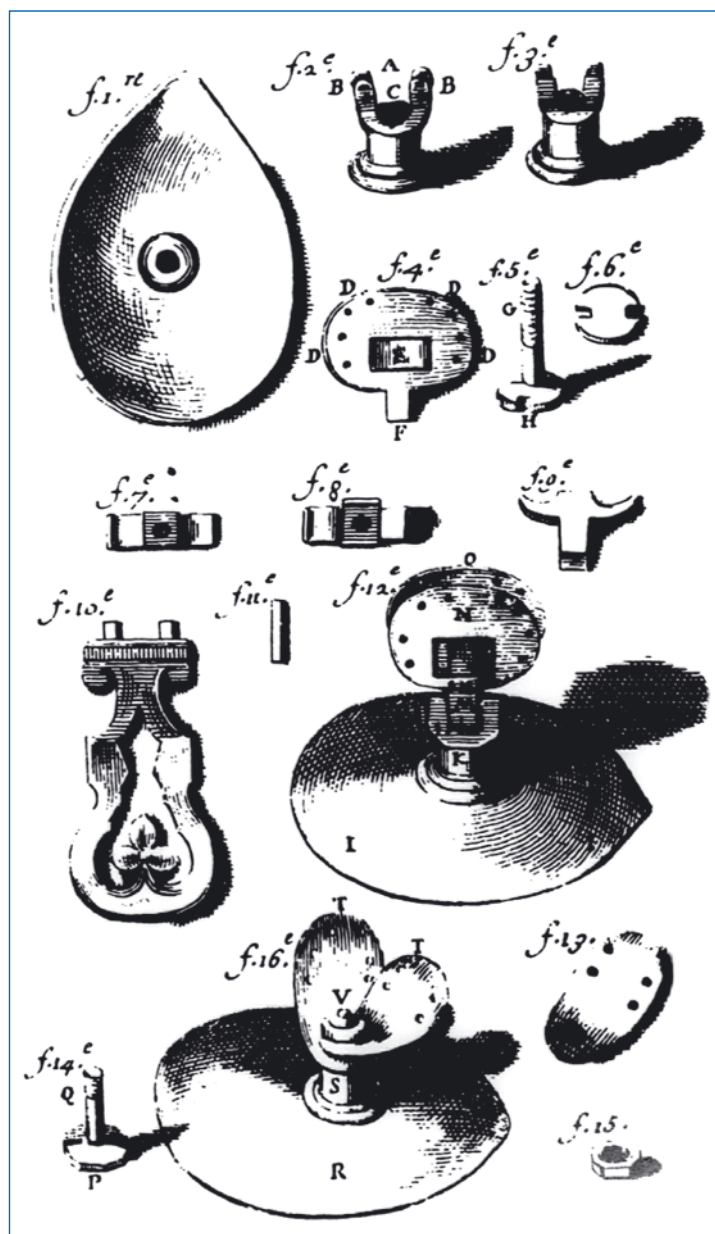


Fig. 38.1. Designs by Fauchard showing early obturators employed for palatal defects. (From [26]. Reprinted in [27])

The decision for prosthetic rehabilitation is made based on the individual patients' needs, motivation for improvement, and availability of the suggested rehabilitative program. Approximately 50% of all patients with cleft lip and palate will need some type of fixed or removable prosthesis by 30 years of age.

As our knowledge and experience in the cleft palate field increased, those of us responsible for providing

prosthetic care recognized the importance of establishing a better prosthodontic concept and principles regarding treatment. In rendering these patients the best service, we should first follow all the rules and principles governing the fixed and removable partial denture prosthesis and, secondly, should remove any fear of causing harm because of existing anatomic, functional, and physiologic deviation.

38.1 Diagnosis and Treatment Planning

In treating people whose oral-facial handicaps affect speech, the best results are achieved when the diagnosis and treatment are carried out by a group of clinicians who represent the various interested specialities and work together as a team rather than independently performing a series of procedures.

In diagnosis and treatment planning, full consideration should be given to: (1) the type and width of the cleft, (2) the position and relation of the maxillary segments to each other in unilateral and bilateral clefts, (3) the form and lateral and anteroposterior dimensions of the maxillary arch, (4) the length, thickness, and mobility of the soft palate, (5) the perforations remaining in the hard and soft palate area and labial sulcus after surgery, (6) the posterior and lateral pharyngeal wall movement and the size of the nasopharynx, (7) a loose premaxilla, (8) the number of missing teeth, (9) malformed and malposed teeth, (10) partially erupted teeth, (11) teeth in the line of the cleft, (12) constricted maxillae, (13) the condition of the tonsils and adenoids, and (14) growth and development of the child. The patient's articulation, voice quality, hearing acuity, mental attitude, and general health also must be considered.

Socially acceptable speech cannot be produced without proper velopharyngeal valving. Therefore, surgical closure of the palate without due consideration of the depth of the nasopharynx and the length and function of the velum during phonation cannot satisfy this objective. Better understanding of the nature of the cleft, anatomy, and the physiology of the area involved would eliminate many of these difficulties. The results of surgical treatment of cleft palates should be evaluated with the aid of cineradiographic studies, nasal endoscopy, serial cephalometrics, maxillary and mandibular casts, speech recordings made before and after surgery, sound spectrographic analysis, measurements of nasal and oral air pressure and flow, and speech and audiometric evaluations.

All members of the team should be thoroughly familiar with the problem at hand. Often the best result is not achieved when the knowledge of the specialists is not all-encompassing [2].

The total habilitation and rehabilitation in the field of oral, facial, and speech impairment is achieved only when the following objectives are kept in mind: (1) socially acceptable speech, (2) restoration of the masticating apparatus, (3) aesthetic facial and dental harmony, and (4) psychological adjustment of the patient to the condition.

Use of a speech appliance simply as a last resort is poor procedure. Its use must be clearly indicated by the oral conditions. For example, the indications for a prosthesis are clearly defined for a patient who has

undergone a series of unsuccessful palatal operations. There is no magic in a prosthetic speech aid. However, there are some patients for whom a prosthesis seems to be the only means of improving speech. In such situations it fills a definite need. A prosthetic speech aid should be used for palatal conditions where it is indicated, just as the pharyngeal flap operation should be used only where it is indicated.

38.2 Treatment Planning

Treatment programs for cleft palate patients require careful planning and should include all factors involved in total health care. The interest of the dentist and physician in craniofacial growth and behavior of soft and hard tissues, both before and after surgery, has increased cooperation between surgeons and dentists. As a result, a dental specialist has the opportunity to examine the cleft palate child, with the surgeon, before any surgery is undertaken. Analysis of longitudinal maxillary and mandibular casts, cephalometrics, and radiographs has shown that two major factors cause growth disturbances of oral-facial regions in individuals with clefts: first, the inherent potential for growth disturbance present among cleft palate patients and, second, the trauma caused by surgical and orthopedic intervention. Because the first factor can be neither predicted nor reduced, efforts have been directed toward minimizing growth disturbance by performing surgery with the least amount of trauma and scar tissue. Longitudinal data obtained during the past 4 years regarding the surgical closure of the cleft with minimum amount of scar tissue and trauma are very encouraging [17].

38.3 Requirements of a Speech Appliance

1. The prosthesis must be designed for the individual patient in relation to his oral and facial balance, masticatory function, and speech.
2. Knowledge related to removable partial and complete dentures should be used in designing the maxillary part of the cleft palate prosthesis. Preservation of remaining dentition and surrounding soft and hard tissue in cleft palate patients is of utmost importance. Improperly designed cleft palate appliances can result in premature loss of both hard and soft tissue, further complicating prosthetic habilitation.
3. The prosthetic speech appliance should have more retention and support than most other restorations. The crowning and splinting of the abutment teeth in adult patients may increase re-

tention and support of the prosthesis and may extend the life expectancy of abutment teeth.

4. Mouth preparations should be completed before making final impressions. In cases where lateral and vertical growth of the maxilla is incomplete and partial eruption of the deciduous and permanent teeth is evident, careful mouth preparations should be made. To provide support of the prosthesis, these preparations may include gingivectomies to expose clinical crowns (to make them usable) and the placement of copings on remaining teeth to prevent decalcification and caries. Osseointegrated implants have been a great help in gaining adequate retention for the prosthesis.
5. The weight and size of the prosthetic speech appliance should be kept to a minimum.
6. The materials used should lend themselves easily to repair, extension, and reduction.
7. Soft tissue displacement in the velar and nasopharyngeal areas by the prosthesis should be avoided.
8. The velar and pharyngeal sections of the prosthesis should never be displaced by movements of the lateral and posterior pharyngeal wall muscles or the tongue during swallowing and speech.
9. The superior portion of the pharyngeal section should be sloped laterally to eliminate the collection of nasal secretions. The inferior portion of the pharyngeal section should be slightly concave to allow freedom of tongue movement.
10. The location and the changes of the speech bulb should include consideration of the following factors:
 - a. The speech bulb should be positioned in the location of greatest posterior and lateral pharyngeal wall activity, because voice quality is judged best when the speech bulb is at these positions.
 - b. The inferior-superior dimension and weight of the speech bulb may be reduced without apparent effect on nasal resonance. (The lateral dimension of the bulb does not change significantly as the position is varied.) (Fig. 38.2)
 - c. The speech bulb should be placed on or above the palatal plane in cases where posterior and lateral pharyngeal wall activities are not present or where visual observation of the bulb is not possible, due to a long, soft palate (Fig. 38.3).
 - d. The anterior tubercle of the atlas bone can be used as a reference point; however, investigation has shown that the relative position of the tubercle of the atlas bone varies in different individuals, and that the positions of the velopharyngeal structures change in relation to the tubercle as the individual moves his or her head.



Fig. 38.2. As a result of our studies, we have concluded that the inferior-superior dimensions of the speech bulb do not have a significant effect on speech quality as long as the bulb is properly placed to facilitate good velopharyngeal closure. This dimension was reduced to one-quarter of its original size, as shown in cast made during fitting for one patient, without apparent effect on nasal resonance

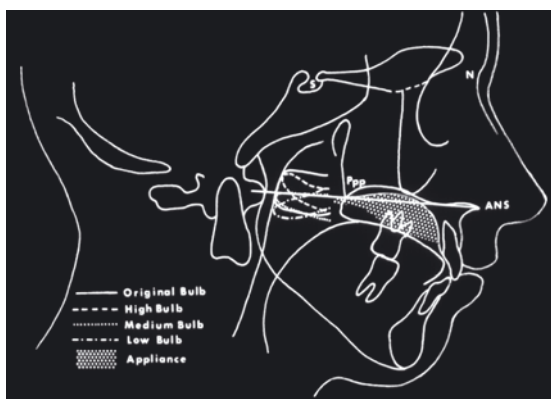


Fig. 38.3. Superimposed tracing of the original speech bulb and various experimental speech bulbs. The palatal plane was used as a plane of reference along with posterior pharyngeal wall activity, muscle bulge, or Passavant's pad. The posterior nasal spine (PNS), absent in cleft palate subjects, is called posterior palatal point (Ppp) and represents the most posterior point of the remnants of the palatal shelves as shown in the lateral cephalometric film. Median position was judged best

Therefore, the atlas bone is no longer used as the reference point for positioning of the pharyngeal section of the bulb.

38.4 Indications for Prostheses in Unoperated Palates

Cleft palate surgery is not a stereotyped exercise, but rather a service that demands an assessment of all factors presented by each patient and a reparative surgical plan based on proven principles. The majority of cleft palates can be reconstructed by surgery, enabling

the patient to develop acceptable velopharyngeal closure. However, in some situations, a prosthesis is the physical restoration of choice. This decision should be made by the group charged with the habilitation of the cleft palate patient.

Many clefts of the hard palate can be closed by a vomer flap [5, 23] and clefts of the soft palate by median suture with good anatomic and functional result. The wide cleft and the short palate demand further attention. Additional length may be gained by a Dorrance or V-Y type retropositioning operation. The raw nasal surface may be covered with a skin graft, nasal mucosa, or an island flap of palatal mucosa [2, 3, 23]. The incompetent palatopharyngeal valve can be augmented by a pharyngeal flap, as either a primary or secondary procedure [24]. The need for additional tissue in a wide cleft can be satisfied by single or double regional flaps.

Despite the surgical advances available to the cleft palate patient, a need remains for cleft palate prostheses. The prosthodontist can assist both the surgeon and patient, and the mutual understanding among the specialists in a well-organized team is of great benefit to the patient. Some situations indicating a prosthetic approach are discussed in the following paragraphs.

38.4.1 A Wide Cleft with a Deficient Soft Palate

Some clefts of this type do not lend themselves to a surgical repair by means of local flaps. A prosthesis is preferable to the more time-consuming remote flaps in these situations. Many patients need a prosthesis to

restore missing dental units, and the distant tissue provides only a dynamic mass (Figs. 38.4, 38.5).

38.4.2 A Wide Cleft of the Hard Palate

In bilateral clefts, the vomer may be high and the cleft of the hard palate wide, so that a surgical repair may produce a low vaulted palate. It may be possible to close the soft palate with the aid of local flaps, and to restore the hard palate with a prosthesis. A situation similar to that once advocated by Gillies and Fry [4] is created: the primary repair of the velum may create a more favorable spatial arrangement for subsequent surgery on the hard palate.

38.4.3 Neuromuscular Deficiency of the Soft Palate and Pharynx

Repair of the palate would not be conducive to the development of good speech. It is difficult to create and maintain a pharyngeal flap large enough to produce competent palatopharyngeal valving without obstructing the airway in the presence of a neurogenic deficiency of the critical muscles. A pharyngeal flap serves best when surrounded by dynamic musculature. When this situation does not exist, the pharyngeal section of a speech-aid prosthesis may serve better to reduce nasality and nasal emission. The prosthesis can also act as a physical therapy modality, providing a resistive mass for the muscles to act against. Should muscle function improve, definitive surgical measures can then be contemplated.

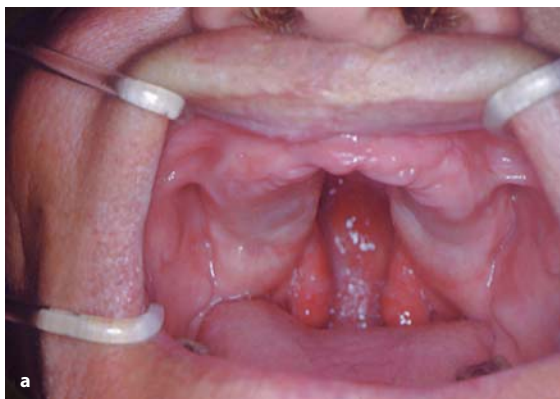


Fig. 38.4. **a** An edentulous patient with an unoperated cleft of the soft and hard palate that affects the retention and support of the prosthesis. At no time should a patient with a cleft, especial-

ly an unoperated cleft, be rendered edentulous. **b** The completed prosthetic speech appliance in position

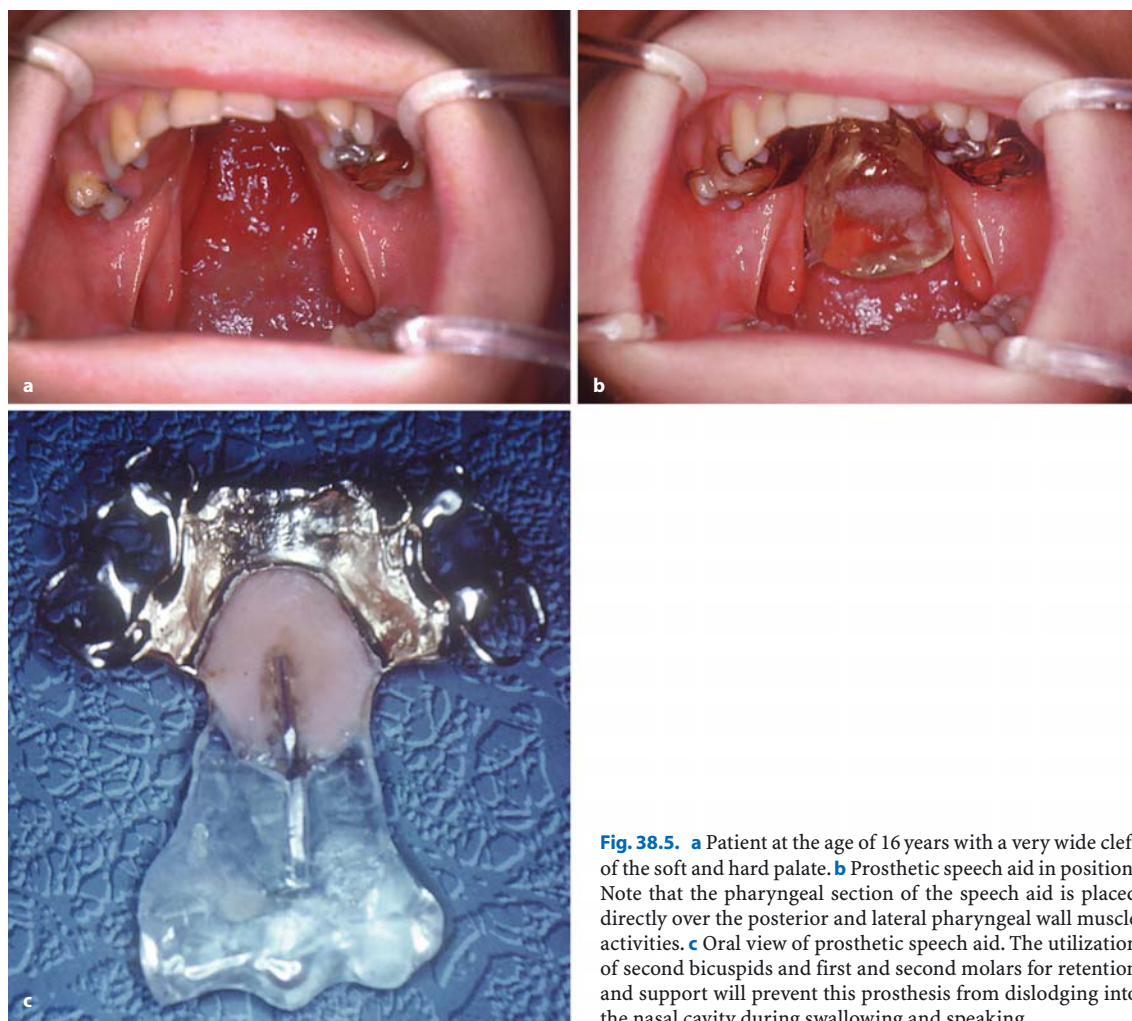


Fig. 38.5. **a** Patient at the age of 16 years with a very wide cleft of the soft and hard palate. **b** Prosthetic speech aid in position. Note that the pharyngeal section of the speech aid is placed directly over the posterior and lateral pharyngeal wall muscle activities. **c** Oral view of prosthetic speech aid. The utilization of second bicusps and first and second molars for retention and support will prevent this prosthesis from dislodging into the nasal cavity during swallowing and speaking

38.4.4 Delayed Surgery

When surgery is delayed for medical reasons, or when the surgeon prefers to repair the palate when the patient is older, the cleft palate may be temporarily closed with a prosthetic speech aid (Fig. 38.6).

38.4.5 Expansion Prosthesis to Improve Spatial Relations

An expansion prosthesis may be used to restore and maintain more normal spatial relations of the maxillary segments prior to surgery. These segments can be gradually separated by an expansion prosthesis to create a space for the premaxilla or to stabilize the parts in a normal position in association with an au-

togenous bone graft. The use of an expansion or repositioning prosthesis, with or without bone grafting, is appropriate for selected cases. In the majority of cleft lip and palate patients, restoration of the anatomic continuity of the labial muscle would mold the segments into acceptable relationships to each other and to the mandible.

38.4.6 Combined Prosthesis and Orthodontic Appliance

An orthodontic appliance may be combined with a prosthesis to move malposed teeth into a more favorable alignment. A prosthetic speech appliance, such as the one illustrated in Fig. 38.7, could be designed for a patient receiving full-band orthodontic treatment.

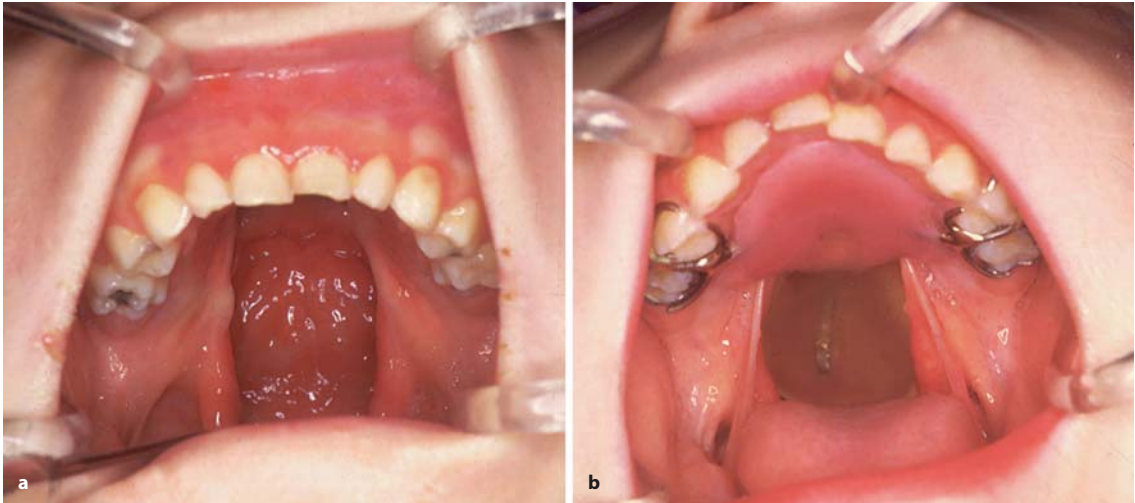


Fig. 38.6. **a** A 4 1/2-year-old girl with a rather wide cleft of the soft and hard palate. We elected to fit her with a prosthesis and to delay the palatal surgery until a later age. **b** The prosthetic

speech aid in position. She tolerated the prosthesis, and the speech significantly improved within a 6-month period

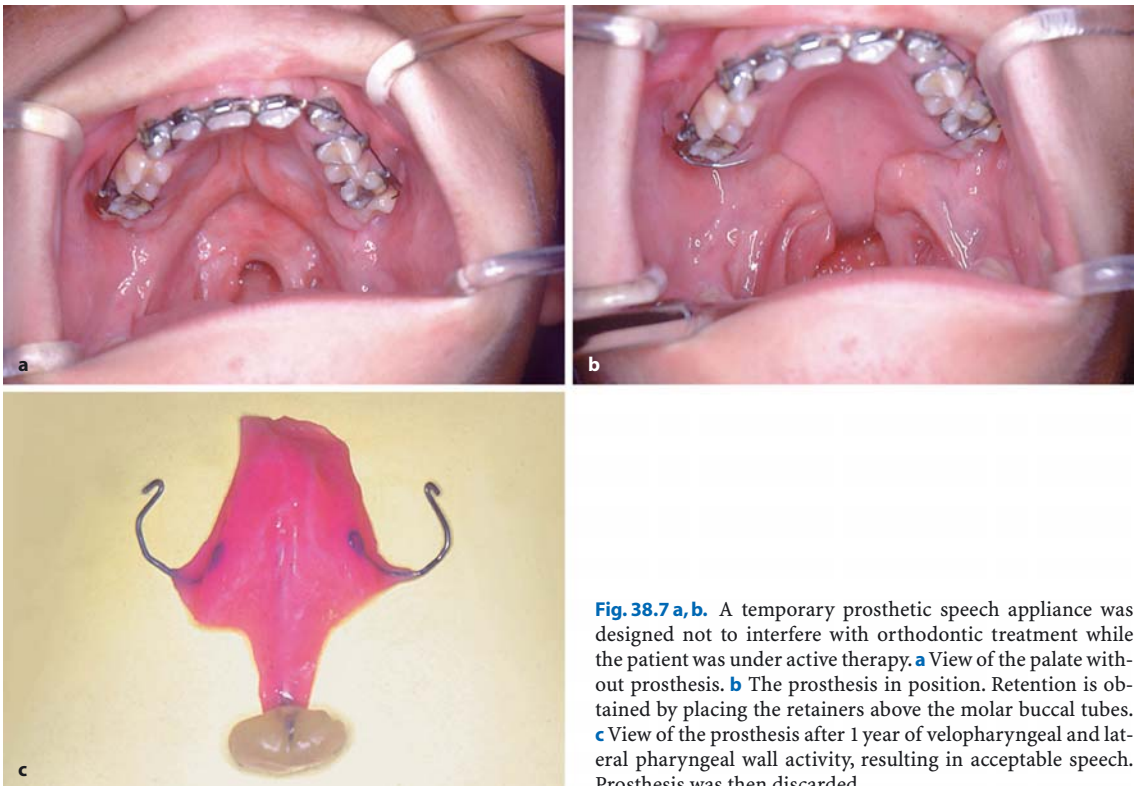


Fig. 38.7 a, b. A temporary prosthetic speech appliance was designed not to interfere with orthodontic treatment while the patient was under active therapy. **a** View of the palate without prosthesis. **b** The prosthesis in position. Retention is obtained by placing the retainers above the molar buccal tubes. **c** View of the prosthesis after 1 year of velopharyngeal and lateral pharyngeal wall activity, resulting in acceptable speech. Prosthesis was then discarded

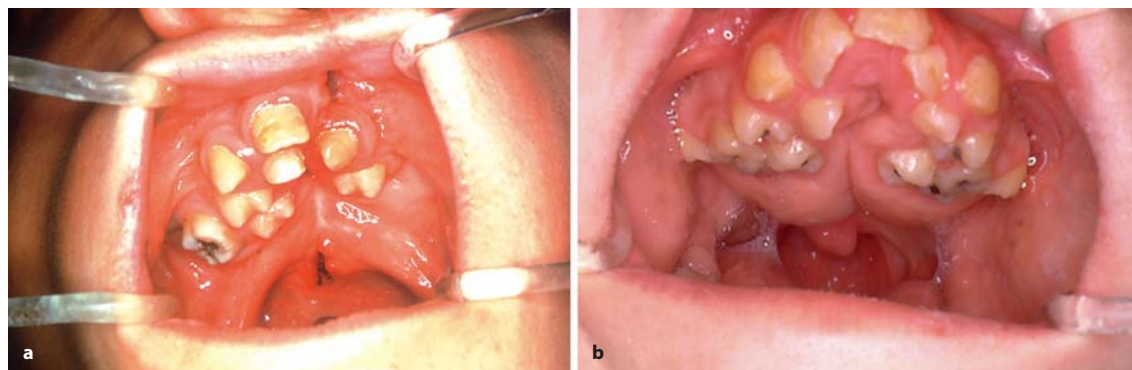


Fig. 38.8 a, b. Two patients with heavily scarred palates and perforations: surgical failures

38.5 Indications for a Prosthesis in Operated Palates

38.5.1 Incompetent Palato-pharyngeal Mechanisms

If clinical, nasal endoscopic, and cineradiographic analyses suggest that the patient is near a functional closure, a prosthesis may serve as a physical therapy modality. The pharyngeal section of the prosthesis is gradually reduced as muscle function improves, and the prosthesis is eventually discarded. When the patient presents a large velopharyngeal gap associated with a neurogenic deficiency, the speech-aid prosthesis should be considered as a permanent treatment.

38.5.2 Surgical Failures

A prosthesis should be considered when a patient presents a low vaulted, heavily scarred, and contracted palate, or a palate with large or multiple perforations (Fig. 38.8). Because of the surgical progress in the last 25 years, plastic surgeons today are not confronted with many failures in cleft palate surgery. Trained surgeons can now predict with greater accuracy the possible success of an operation, and are likely to avoid failure because other alternatives are available. Approximately 50% of all cleft palate patients will need some type of prosthesis by the age of thirty.

38.6 Contraindications for a Prosthesis

1. Surgical repair is feasible only when surgical closure of the cleft will produce anatomic and functional repair.
2. Patients with mental retardation are not good candidates for prostheses, because they frequently are not capable of giving the appliance the care it requires.
3. A speech aid is not recommended for an uncooperative patient, or for a child with uncooperative parents.
4. If caries are rampant and not controlled, a prosthesis will require unusual care, and frequent examinations are important.
5. The edentulous condition is not a contraindication for a speech-aid prosthesis.
6. Because the construction of a functional prosthesis requires the services of a dentist who has had training in cleft palate prosthodontics, it would be better to resort to surgical ingenuity when experienced prosthodontic help is not available.

38.7 Constructing Prosthetic Speech Appliances

For patients with deciduous, mixed, or permanent dentitions that are not fully erupted, all three sections of the prosthetic speech appliance are made of acrylic resin, and wrought wire retainers are used (Fig. 38.9). In patients whose permanent teeth are fully erupted, the anterior section of the prosthetic speech appliance should be made of cast metal or a combination of cast metal and acrylic resin (Fig. 38.10).



Fig. 38.9. A temporary acrylic resin speech appliance with wrought wire clasps and full palatal coverage designed for a 4-year-old child



Fig. 38.10. A permanent cast gold speech appliance with partial palatal coverage for an adult with no missing teeth

38.7.1 Preliminary Impression

A stock tray of adequate dimensions is selected. If a registration of the entire cleft is desirable, the stock tray is modified with modeling compound extending posteriorly to the postpharyngeal wall. The added section is underextended about 4–5 mm in all directions, leaving adequate space for impression material. Fast-setting, irreversible hydrocolloid is used for registering the preliminary impression. The following suggestions should be kept in mind when the preliminary impression is made:

1. If the patient is a child, he or she should be given the opportunity to examine the tray; in some cases the child may be permitted to try the tray in his mouth. Children should be told that their cooperation is needed; otherwise, it will be necessary to make several impressions. Talking to children throughout the procedure is helpful.
2. The patient should have an early morning appointment.
3. The patient should have an empty stomach.
4. A topical anesthetic should be used on a child who has a severe gag reflex.
5. The tray should not be overloaded with impression material. Excess material in the nasopharynx will increase the difficulty of removing the impression without a fracture (see Fig. 38.14).
6. All oral perforations should be packed with gauze that has been saturated with petroleum jelly.

38.7.2 Preparation of the Deciduous Teeth for Retention

Most deciduous teeth do not have sufficient undercut for retention of the prosthesis. However, a small amount of bilateral undercut can give adequate retention. The following recommendations will help to produce adequate retention:

1. Carefully extend the clasp arms into interproximal areas of the teeth.
2. Insert, if necessary, serrated platinum pins into the buccal surface of deciduous molars to create an artificial undercut for the clasp.
3. Place bands with soldered retention lugs on the teeth.
4. Use chrome-cobalt crowns with retention lugs for teeth with extensive carious lesions or areas of decalcification.

After the clasp design has been determined on the diagnostic casts and the teeth have been prepared for retention, the final impression is made. If adequate retention is not available in the permanent dentition, crowning of the molars might be desirable to provide proper retentive areas (Figs. 38.11, 38.12).

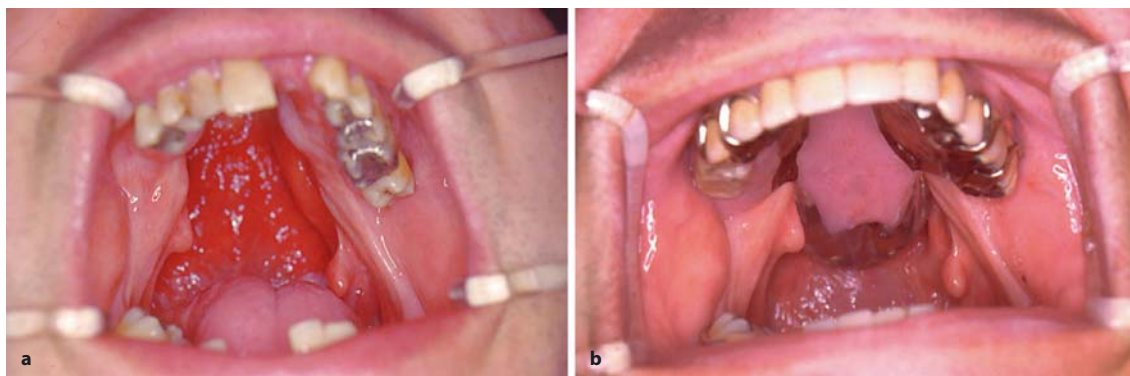


Fig. 38.11 a,b. Crowning and splinting of the abutment teeth will increase the retention and support of the prosthesis and the life expectancy of the abutment teeth. **a** Patient before dental restoration. **b** After restoration with prosthesis in position



Fig. 38.12. Patient with wide cleft of the hard and soft palate, treated with prosthetic speech appliance



Fig. 38.13. An acrylic tray is made over the diagnostic cast and the border trimmed with green modeling compound

38.7.3 Final Impression

An acrylic resin tray is constructed over the diagnostic cast (Fig. 38.13). The patient is prepared in the same manner as for the preliminary impression, and the final impression is then made with an irreversible hydrocolloid impression material (Fig. 38.14). The master cast is made of dental stone.

38.7.4 Jaw Relation Records

Jaw relation records such as vertical dimension, centric relation, and protrusive relation are made and used in the adjustment of the articulator.



Fig. 38.14. The final impression is made with alginate material. Note the extent of the registration of the cleft

38.8 Design and Construction of the Prosthesis

The master casts are surveyed and the prosthesis is designed (Fig. 38.15). For patients with severely constricted maxillary and mandibular arches, teeth are arranged outside the remaining natural teeth to establish the proper aesthetics and occlusion.

The prosthetic speech appliance is constructed in three sections. The design of the anterior portion is similar to that of a partial or complete denture. After this section is completed, the patient is instructed to wear it for at least 1 week. The length of this adjustment period depends on the ability of the patient to adapt to this part of the prosthesis. The construction of the middle part, the tailpiece or velar section, varies for operated and nonoperated clefts.

In unoperated clefts with the maxillary prosthesis in position, the extent of the tailpiece over the margin of the cleft is marked on the posterior part of the appliance. The tailpiece extends posteriorly to the anterior extent of the uvula.

In operated palates that are short and require a prosthesis, the position of the tailpiece is marked on the posterior margin of the prosthesis. The tailpiece extends approximately 3 mm behind the posterior margin of the soft palate. The width of the tailpiece is approximately 5 mm, and its reinforced thickness is about 1.5 mm.

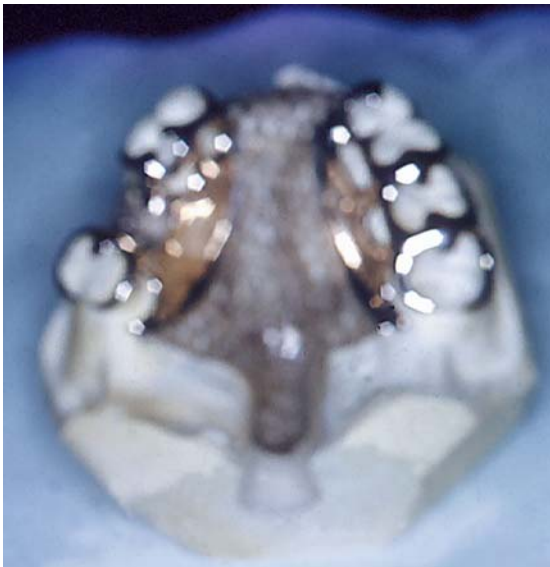


Fig. 38.15. Cast gold framework. The prosthetic speech appliance requires more retention and support; therefore, all the remaining maxillary teeth of this patient have been used for this purpose. The posterior extension of the framework reinforces the tailpiece and the speech bulb

38.8.1 Construction of Velar Section

A piece of shellac baseplate material of the required width and length is used as a tray. It is securely attached to the posterior part of the prosthesis with about 2-mm relief. This assemblage is examined in the patient's mouth for proper extension. The tissue side of the tray is filled with zinc oxide and eugenol impression paste, and the appliance is inserted into the mouth. The patient is instructed to hold his or her head in a vertical position to prevent escape of the impression material into the nasopharynx. The head is held in this position for 1 minute, then the patient is instructed to swallow a little water so that the muscular movement of the soft palate will be registered in the impression. After the material has hardened, the prosthesis is removed from the mouth, and the tailpiece is processed with self-curing acrylic resin. The denture portion with the finished tailpiece is placed in the mouth for testing. Swallowing of small amounts of water will stimulate muscle action along the lateral edge of the velar section. If the velar section is overextended laterally, undue muscle displacement and eventual tissue soreness will occur.

38.8.2 Construction of Pharyngeal Section or Speech Bulb

Two holes are drilled in the posterior part of the tailpiece. A piece of separating wire is drawn through the holes to form a loop that extends superoposteriorly beyond the superior part of the tailpiece. The ends of the wire are twisted together inferiorly (oral side), and secured to the appliance by sticky wax (Fig. 38.16). The wire loop that is extended into the nasal pharyngeal area is manipulated into an oval form, and the appliance is inserted into the mouth (Fig. 38.17). The patient is asked to swallow, and the wire is adjusted so that it will not contact the pharyngeal walls at any time. Posterior and lateral pharyngeal wall activity can be stimulated by spraying those tissues with water. The desired position of the wire is in the area of the maximum posterior and lateral pharyngeal constriction. Green modeling compound is added around the wire loop to reinforce it and its attachment to the tailpiece (Fig. 38.18). The appliance is inserted into the patient's mouth, and he is asked to swallow a little water. Adaptol, softened in water at 150° to 160°F for 4–5 min, is added over the green compound, and the appliance is inserted into the mouth. Again the patient is instructed to swallow a little water to produce muscle activity, and thus the impression material is molded (Fig. 38.19).

The prosthesis is reinserted a number of times, and the patient is instructed to swallow each time when



Fig. 38.16. **a** The location of the two holes drilled on the tailpiece. **b** View of the wire formed in a loop, extending superoposteriorly into the nasal pharynx



Fig. 38.17. Wire loop is attached to the tailpiece, inserted into the mouth of patient seen in Fig. 38.12, and checked to see that it does not contact posterior and lateral pharyngeal walls during swallowing



Fig. 38.18. Modeling compound is added around the wire loop to reinforce the wire and its attachment to the tailpiece



Fig. 38.19. Adaptol, softened by heating to 150° to 160° F, is added over the green compound, and appliance is inserted into the mouth of the patient, Fig. 17.12. Note the displacement of the material after patient has swallowed some water and rotated the head to each side and down

additions of Adaptol are made to the mass on the wire loop. These steps are repeated until a functional impression of the lateral and posterior pharyngeal walls is made (Fig. 38.20). The impression material is then molded by instructing the patient to place his chin against his chest and move his head from side to side. In the rest position, he swallows water and talks to allow further molding of the impression material by muscular activity. If the mass is overextended, the patient will feel it during these actions. The overextended bulb impression is easily adjusted by reheating the bulb on the exterior surface and reinserting it into the

patient's mouth. While the material is soft, the patient is instructed to produce the desired muscular activities. The completed speech bulb impression is chilled thoroughly in ice water. To check the position of the bulb, water is injected again, and the position of the bulb is examined in the mouth for its relation to the posterior and lateral pharyngeal wall activities. A spray of water onto the tissue will again stimulate these activities. In unoperated clefts, muscle function along the speech bulb during swallowing can be observed directly when the mouth is wide open and water is being injected onto the tissues. When the posterior pharyngeal wall activity is not present, or direct visualization is not possible due to the length of the soft palate, a lateral cephalometric radiograph will reveal the position of the bulb in relation to the nasopharyngeal structures. In such cases, the bulb is placed in the area of the palatal plane. When the bulb form has been perfected, the bulb and tailpiece are processed onto the denture portion of the appliance. A heat-cured acrylic resin is used for making these parts.

For patients with unusually sensitive posterior and lateral pharyngeal walls (e.g., when the gag reflex is easily triggered), the making of a final impression for the speech bulb on the initial try is delayed until the patient is properly prepared for the impression. In such cases, it is helpful to construct an underextended bulb in self-curing acrylic resin, and to allow the patient to become adjusted to this small bulb for 2 or 3 weeks. After the patient has become accustomed to the undersized bulb, a final impression is made by adding Adaptol to the bulb, following the procedures



Fig. 38.20. **a** Functional registration of the velopharyngeal region using Adaptol. The gradual addition of Adaptol and patient swallowing water and moving the head down and to the sides will give the functional impression of the velopharyngeal region. If any gagging reflex is present, then underextended pharyngeal section is processed using an autopolymer. A week or two later the pharyngeal section is modified for addition of

the Adaptol. When the desired speech result is obtained and the patient does not show any gagging reflex, the speech bulb is heat-processed. **b** More Adaptol is gradually added, and the appliance is inserted until a functional impression of the area has been obtained. In most patients, the speech bulb does not contact the throat wall while the surrounding tissues are at rest



Fig. 38.21. Processed speech bulb in position, patient from Fig. 38.12



Fig. 38.22. The nasal and lateral sides of the speech bulb, tailpiece, and a portion of the palatal area of the anterior section are placed in dental stone. These parts of the appliance are made of acrylic resin

previously outlined. The final impression of the speech bulb is processed in a heat-curing type of acrylic resin (Figs. 38.21, 38.22).

To prevent the patient from swallowing the bulb in case the tailpiece is fractured, the appliance should be reinforced by incorporating a piece of No. 11 gauge half-round wire in the anterior body of the appliance and extending the wire into the bulb. If the anterior part of the appliance is made of cast metal, the frame should be extended posteriorly to strengthen the velar and pharyngeal section (see Fig. 38.15).

38.8.3 Insertion of the Appliance

The finished speech appliance is inserted into the mouth and examined for muscle adaptation to the speech bulb during swallowing and phonation, excessive pressure against the posterior and lateral walls of the pharynx, stability of the appliance during function, and improvement of the quality of the voice.

38.8.4 Position of Speech Bulb

For most patients, when the bulb is positioned too far inferiorly, the pharyngeal section has the following undesirable effects:

1. It has a tendency to be displaced by the dorsal part of the tongue during tongue movements.
2. It fails to relate to the normal region for making adequate velopharyngeal closure.
3. It has a detrimental acoustical effect on the quality of the voice. (Caution should be exercised to avoid blocking or extending the speech bulb into the eustachian tube.)

38.9 Summary

The prosthetic treatment of certain patients with cleft palate is an important part of the multidisciplinary approach to solving the many problems related to total health.

Some of the cleft palate patients for whom speech aids can be made include those with a wide cleft of the palate with a deficiency of the soft palate, a wide cleft of the hard palate with a high vomer, a neuromuscular deficit (a sphincteric velopharyngeal action may not be attained even with a pharyngoplasty if the deficit is marked), and surgical failures.

I strongly object to the use of remote extraoral flaps in cleft palate surgery, because a prosthesis seems to be more appropriate. The possibility of cancer being related to such a prosthesis is quite remote, and there has been no evidence of increased hearing loss in patients wearing a prosthesis. A prosthesis should not be used in a patient not competent to care for it or maintain proper hygiene.

A prosthodontist engaged in treating patients with oral, facial, and speech deficits should be thoroughly familiar with the anatomic and physiologic deviations of the region involved and with the basic principles involved in prosthetic dentistry. He should always be willing to acquire further knowledge in this field.

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39 Palatal Lift Prosthesis for the Treatment of Velopharyngeal Incompetency and Insufficiency

MOHAMMED MAZAHERI

39.1 Treatment, Methodology, and Results in Patients with Velopharyngeal Inadequacy

Before getting into methodology of treatment of patients with velopharyngeal inadequacy, who require prosthetic velar elevation and velopharyngeal stimulation, let us outline the Lancaster Cleft Palate Clinic's present concept of treatment for patients with various types of velopharyngeal incompetency.

From 1984 to 1992, a total of 431 patients were referred to the Lancaster Cleft Palate Clinic with congenital or acquired velopharyngeal incompetency (VPI) (Table 39.1). This population consisted of 230 males and 201 females with a mean age of 11.26 years. Note the breakdown in the type of velopharyngeal incompetency. Two hundred seventy-one patients (63%) demonstrated congenital velopharyngeal incompetency without submucous cleft; 86 (20%) had VPI with a submucous cleft; 68 (16%) had VPI related to trauma; and 6 (1%) had VPI as a result of diseases such as myasthenia gravis, stroke, polio, and other neurological disorders.

Each patient was examined and evaluated by a plastic surgeon, prosthodontist, and speech-language pathologist with a combined experience of 110 years. A questionnaire was designed for data acquisition and long-term follow-up of these patients (Table 39.2).

39.1.1 The Referral

It is interesting to note that 256 patients (59%) were referred by speech-language pathologists (Table 38.3), indicating that velopharyngeal incompetency is not recognized at an early age and that the diagnosis is frequently made when the patient starts school. The number of physician referrals was 96 (22%). The remaining referrals (19%) came from rehabilitation counselors, dentists, rehabilitation centers, and families.

Table 39.1. Number of patients and type of velopharyngeal incompetency (VPI)

	Number of Patients
Male	230 (53%)
Female	201 (47%)
	Type of Velopharyngeal Incompetence
No cleft	271 (63%)
Submucous cleft	86 (20%)
From trauma	68 (16%)
From disease	6 (1%)

Please note that 104 patients (25%) had had their tonsils and adenoids removed in order to eliminate or remedy the velopharyngeal incompetency (Table 39.4). This, of course, causes an increase in hypernasality for the VPI patient.

In addition to oral examination, nasal endoscopy, and individual judgment, all patients had two cephalometric radiographs taken, one with the soft palate at rest and the second during prolonged phonation of the vowel "E." Twenty-five percent of the subjects had cineradiographic studies of the velopharyngeal region to observe continuous phonation.

39.1.2 Results of Treatment

According to nasal endoscopic evaluation, radiographic analysis, and listener judgments, 126 (29%) of the patients demonstrated inconsistent or borderline velopharyngeal dysfunction (Table 39.5). The team decided that each subject was to be referred to a speech-language pathologist with instructions to have a review by the team in 1 year if the condition

Table 39.2. Questionnaire designed to record appropriate information on patients for the study

1. Patient number _____

2. Patient name _____

3. Patient address _____

4. Birth date _____ 5. Sex _____ Race: _____

6. Referral source _____

7. Chief complaint _____

8. Specific diagnosis:

 Congenital VPI, no cleft _____

 VPI with cleft _____

 VPI with submucous cleft _____

 VPI from trauma _____

 VPI from cancer _____

 VPI with other diseases _____

 Iatrogenic VPI _____

 Other unclassifiable _____

9. Diagnostic data available

 Cephs _____

 Lateral only _____

 Lateral and AP _____

 At ages _____

 No cephs _____ Tracings _____

 Cineradiographs

 Yes _____ at ages _____

 No _____

 Recordings

 Yes _____ at ages _____

 No _____

 Dental models

 Yes _____ at ages _____

 No _____

 Growth analysis

 Yes _____ No _____

 Sonagrams

 Yes _____ No _____

 Audiology Examination

 Yes _____ at ages _____

 No _____

 Surgical records

 Yes _____ No _____

10. Other conditions (Short narrative or diagnostic classification)

 Dental health: _____

 Orthodontic: _____

 Audiology, otology: _____

 Allergies: _____

 Smoking habits:

 No _____ Yes _____ Pack/day _____

 Tonsils and adenoids removed:

 Yes _____ age _____

 No _____

Table 39.2. (Continued)

11. When was VPI first noticed:	
Age of onset	_____
Circumstance	_____
Who first noted VPI	_____
12. VIP treatment history	
Speech therapy _____	No. of sessions _____
Surgery (flap) _____	Type of flap _____
Surgeon _____	Hospital _____
Other surgery _____	Procedure _____
Surgeon _____	Hospital _____
Prosthesis _____	Type of prosthesis _____
Prosthodontist _____	Hospital/clinic _____
13. Sequence of treatment (if multiple procedures)	
Speech therapy only	_____
Flap and speech	_____
Flap only	_____
Lift and speech	_____
Lift only	_____
Speech and flap	_____
Speech and lift	_____
Flap and lift	_____
Lift and flap	_____
Three procedures sequence:	
1. _____	2. _____ 3. _____
14. Evaluation of result (speech)	
Date of last follow-up	_____
Acceptable	_____
Not acceptable	_____
Acceptable but can improve	_____
Not acceptable but can improve	_____
No improvement likely	_____
Should recall patient	_____
15. Recommendations	
Today's date _____ Preparer's signature _____	

persisted. Further evaluation of these patients after 1 year revealed that the hypernasality or nasal emission had subsided, and none required further treatment.

It was recommended that 177 patients (41%) have pharyngeal flap surgery. In 122 patients (mean age, 10 years), the surgical procedure consisted of a superiorly based flap performed by our staff plastic surgeon. The remaining 55 subjects were referred to the plastic surgeon of their choice for a pharyngeal flap with instructions to return to the Clinic after insertion of the flap for further evaluation.

Thirteen of the subjects with congenital VPI who were treated with a pharyngeal flap continued to exhibit a significant to moderate amount of hypernasal resonance and nasal emission (Tables 39.6, 39.7). Palatal lifts or combination prostheses were constructed for these patients. Five of these patients had the palatal lifts removed, and two had their combination appliance removed after 1 year because the prostheses had resulted in their developing adequate posterior and lateral pharyngeal wall activity, and the patients were judged to have satisfactory voice quality without the appliances. Five of the patients with a lift

Table 39.3. Referral source for VPI Patients

Source	No. of Referrals	Percent of Sample
Speech pathologist	256	(59%)
Physician/surgeon	98	(23%)
RN (nurse)	23	(5%)
Dentist	12	(3%)
PDH/BVR	23	(5%)
Rehabilitation center	1	(.5%)
Social worker	2	(.5%)
Family	18	(4%)

Table 39.4. Status of tonsils and adenoids of VPI patients

Status of Adenoids	No. of Cases	Percent of Sample
In	100	(23%)
Out	104	(24%)
In/Out (to insert pharyngeal flap)	14	(3%)
No information available	213	(50%)

and one with a combination appliance continued wearing their prostheses because of consistent nasal emission and lack of response to the prosthetic stimulation. One patient with VPI as a result of trauma who had pharyngeal flap surgery continued wearing his combination prosthesis. The remaining patients with pharyngeal flaps were judged to have acceptable speech quality by the three team members. Further tests for nasal and oral pressure (cul-de-sac shifting, listening tube, nasal endoscopy, and oral manometer) substantiated the clinical finding.

Eighty-nine of the subjects were fitted with a palatal lift or combination prosthesis (Table 38.8). Sixty-one of the patients with congenital VPI (mean age, 11 years) had a palatal lift or combination appliance. At the time of this study, 13 of the patients with a palatal lift were still wearing their prostheses and 21 had gained adequate muscle activity so that the prostheses were removed. Twenty-three of the 61 patients were still wearing a combination lift, and four gained adequate tissue stimulation, so the prosthesis was discarded.

Of the 19 patients with traumatic VPI (mean age, 21 years), 11 had their prostheses still in position, four were removed, one had his combination in position, and three had rejected the combination prosthesis

Table 39.5. Treatment methodology for patients with VPI

Treatment	No. of Cases	Percentage of Sample
Speech only	126	(29%)
Pharyngeal flap	122	(28%)
Pharyngeal flap recommended	55	(13%)
Palatal lift	74	(17%)
Palatal lift removed after stimulation	8	(2%)
Palatal lift recommended	15	(3%)
Pharyngeal flap – palatal lift	16	(4%)
Palatal lift – pharyngeal flap	12	(4%)

Table 39.6. Status of patients who received palatal Lifts

Status	No. of Patients
Palatal lift appliance removed for pharyngeal flap	3
Pharyngeal flap patients received appliances	13
Lift removed later	5
Combined appliance removed later	5
Still wearing palatal lift	5
Combined appliance still being worn	1

Table 39.7. Summary of use of prostheses and pharyngeal flaps

Pathology	Palatal lift			Combined appliance	
	N	In	Out	In	Out
Congenital					
PF-PL	9	4	5	0	0
PF-COMB	3	0	0	1	2
PL-PF	3	0	3	0	0
Trauma					
PF-PL	1	1	0	0	0
Totals					
Appliance in	6	5	0	1	0
Appliance out	10	0	8	0	2

Note: PF = pharyngeal flap, PL = palatal lift prosthesis, COMB = combination prosthesis.

because of difficulty of adjustment, more difficult swallowing, or lack of patient motivation and/or co-operation.

Of the nine patients with VPI as a result of various neurological diseases, three have a palatal lift in posi-

Table 39.8. Summary of status of palatal lift and combination prostheses used for VPI of various etiologies

Etiology	Palatal lift			Coinhination	
	N	In Position	Removed	In Position	Removed
Congenital	61	13	21	23	4
Trauma	19	11	4	1	3
Disease	9	3	3	3	0
Totals	89	27 +	28 = (55)	27 +	7 = (34)

Note: Of the 35 appliances that were removed, 3 were removed due to patient rejection and 32 were removed after increased velopharyngeal function and satisfactory speech quality were achieved.

tion, three appliances have been removed, and three have a combination appliance still in position.

Fifteen additional patients were recommended for palatal lift prostheses, but the subjects or subjects' families elected not to have any form of treatment. Six-month follow-up revealed that the patients or the parents were satisfied with the patient's speech as it was. It was recommended to five patients that their palatal lift be removed in favor of a pharyngeal flap performed by a plastic surgeon in the patient's hometown. There was no follow-up at the Clinic for these patients after the insertion of the flap.

39.1.3 Summary

Analysis of the 35 patients whose appliances were removed revealed that three patients rejected the prosthesis within 6 months. Of the remaining 32 patients, three appliances were removed to insert a superiorly based pharyngeal flap, and 29 were removed when the patient demonstrated voice quality without the appliance that was judged to be satisfactory. Hypernasality was no longer a concern to these patients. The judges found this to be accurate.

In our population, use of the palatal lift or combination appliance for patients with traumatic VPI resulted in more acceptable speech performance than with velopharyngeal flap.

39.1.4 Conclusion

It is interesting to note that a majority of the patients referred to the Lancaster Cleft Palate Clinic for velopharyngeal incompetency were referred by speech-language pathologists. It was also interesting to note that a significant number of patients had had their tonsils and adenoids removed to remedy their hypernasality.

We have found that patients with a gap of more than 12 millimeters between the soft palate and poste-

rior pharyngeal wall respond more favorably to physical therapy with a palatal lift or combination prosthesis prior to a pharyngeal flap than patients who have a pharyngeal flap as the initial mode of treatment.

Two of the subjects with complete paralysis of the soft palate as a result of traumatic injury had pharyngeal flaps performed by non-team member plastic surgeons; neither of these surgeries produced an acceptable speech quality result.

It is also interesting to note that a majority of the patients were diagnosed as having velopharyngeal incompetency after the age of 5. The studies show that the younger patients responded much more favorably to our treatment modalities (pharyngeal flap, palatal lift) than older patients. Therefore, it behooves us to diagnose cases at earlier ages and undertake the required treatment as early as possible.

39.2 Palatal Lift Prostheses for the Treatment of Patients Requiring Velar Elevation, Velopharyngeal Stimulation, and Velopharyngeal Obturation

39.2.1 Symptoms

Hypernasality or nasal emission and decreased speech intelligibility occur as a result of several organic conditions, (e.g., congenital or acquired cleft of the palate, congenital short soft palate or palatal paresis or velopharyngeal insufficiency, velar paralysis or velopharyngeal incompetency, abnormal nasal pharyngeal size, and hypernasality occurring after the removal of the tonsils and adenoids).

39.2.2 Etiology

The etiological factors contributing to the development of these organic conditions can be classified into two major categories:

1. Prenatal
 - a. Cleft of the palate
 - b. Short soft palate
 - c. Abnormal nasal pharyngeal size
 - d. Abnormal velopharyngeal neuromuscular development
2. Postnatal

Partial or completely paralyzed velum as a result of central or peripheral nervous system damage (e.g., a patient with myasthenia gravis, bulbar polio, traumatic brain injuries, cerebral vascular accidents, degenerative central nervous system diseases, and amyotrophic lateral sclerosis).

39.2.3 Speech Characteristics

Speech characteristics common in both types of patients with velopharyngeal incompetency and velopharyngeal insufficiency are:

1. Hypernasality
2. Nasal emission
3. Decreased intelligibility of speech due to weak consonant production

The patient with velopharyngeal insufficiency often develops glottal stop substitution as a result of compensation for production of pressure consonants. The patient with neurological diseases resulting in a full or partial paralysis of lips, tongue, larynx, or respiratory musculatures often develops an abnormal articulatory pattern and diminution of breath pressure, which causes a reduction of oral pressure and flow.

39.2.4 Methods of Treatment

The closure and obturation of palatal clefts and defects for patients with congenital and acquired clefts have been reported. Early humans used stone, wood, gum, cotton, and other foreign bodies to obturate the palatal opening. In recent years, several methods have been advocated for satisfying the main objective of socially acceptable speech for these patients. Among these concepts are:

1. Traditional speech treatment, such as active lip, tongue, and palate exercises for the stimulation and physical therapy of musculatures (myofunctional therapy), designed to effect reduction in hypernasality.
2. Surgical methods designed to reduce the velopharyngeal gap or lumen, employing velar lengthening procedures, velopharyngeal flaps, implants (cartilage, bone, silicone, Teflon®), and combinations of several methods.

3. Faradization and electrical vibration massage to stimulate palatal function.
4. Prosthetics designed to elevate and stimulate the soft palate in patients with velopharyngeal incompetency or to elevate, stimulate, and obturate the velopharyngeal lumen in patients with velopharyngeal insufficiency.

As previously stated, two prosthodontic procedures are available to us in the treatment of patients with velopharyngeal inadequacies:

1. Lift type
2. Combination of lift and bulb

The lift type of prosthesis is used to elevate the soft palate to the maximum position attained during normal speech and deglutition. The reduction in size of the velopharyngeal gap and lumen will decrease nasal air flow, increase oral pressure for consonant articulation, and improve voice quality. The lift may also act as a physical modality for stimulation of velar and pharyngeal musculatures and elimination of the occurrence of velar disuse atrophy (Figs. 39.1–39.6).

The combined lift/bulb prosthesis should be the method of choice when the soft palate is insufficient for the proper velopharyngeal closure. The combined lift/bulb prosthesis is used to elevate the soft palate, obturate the gap, and stimulate velopharyngeal development and pharyngeal constriction (Figs. 39.7, 39.8).

39.2.5 Prerequisites of Lift and Combination Prostheses

1. The maxillary portion of the prosthesis is designed to achieve optimal retention and stability.
2. The lift portion should be placed so that velar elevation occurs in the area where normal velopharyngeal closure takes place.
3. Elevation of the velum should be gradual so that the velum becomes less resistant to displacement.
4. The pharyngeal section should be placed in the area where posterior and lateral pharyngeal constriction takes place so that it increases the change of further stimulation and muscle activation.
5. The reduction of pharyngeal section, when indicated, should be gradual.
6. Speech therapy, such as active lip, tongue, and palatal exercises and placement, should be properly instituted in conjunction with the construction and insertion of the prosthesis.

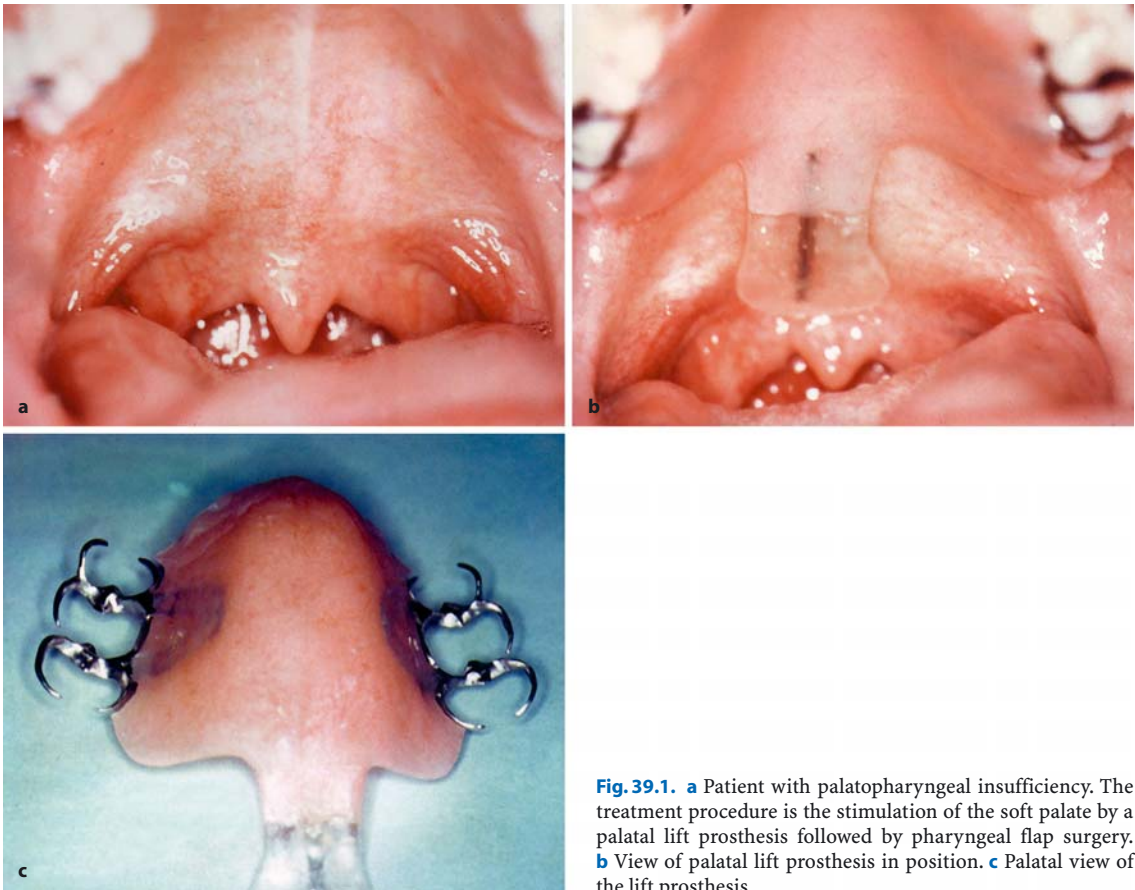


Fig. 39.1. **a** Patient with palatopharyngeal insufficiency. The treatment procedure is the stimulation of the soft palate by a palatal lift prosthesis followed by pharyngeal flap surgery. **b** View of palatal lift prosthesis in position. **c** Palatal view of the lift prosthesis

39.2.6 Objectives in Making Prosthetic Lift and Combination Services

1. Reduce hypernasality and nasal air escape by velar elevation
2. Reduce the degree of disuse atrophy
3. Increase velopharyngeal function by constant and continuous stimulation
4. Increase neuromuscular response by gentle stimulation and speech exercises

39.2.6.1 Results of Using Lift and Combination Prostheses

Methods of Evaluation

1. Speech testing procedures
2. Nasal endoscopy
3. Radiographic evaluation (e.g., cineradiography, cephalometrics, sectional laminography, or tomography)

4. Oral nasal air pressure and air flow assessing devices
5. Electronic instrumentation such as Tonar and sonograph

The optimal result depends on the type of oral pharyngeal involvement. If the neurological disorder is more localized to the velopharyngeal region, and the patient has few or no speech articulatory disorders, the prosthetic result is optimal. Patients with muscle paralysis of the tongue, lips, larynx, and respiratory organs usually respond less favorably to prosthetic care. Their phonatory and articulatory disorders usually remain after the prosthetic treatment. These patients often require more intensive and coordinated myofunctional therapy.

Patients' tolerance and acceptance of prosthetic treatment vary. Some patients have less difficulty than others, becoming accustomed to the palatal and velopharyngeal coverage and decreased oral pharyngeal space and volume.

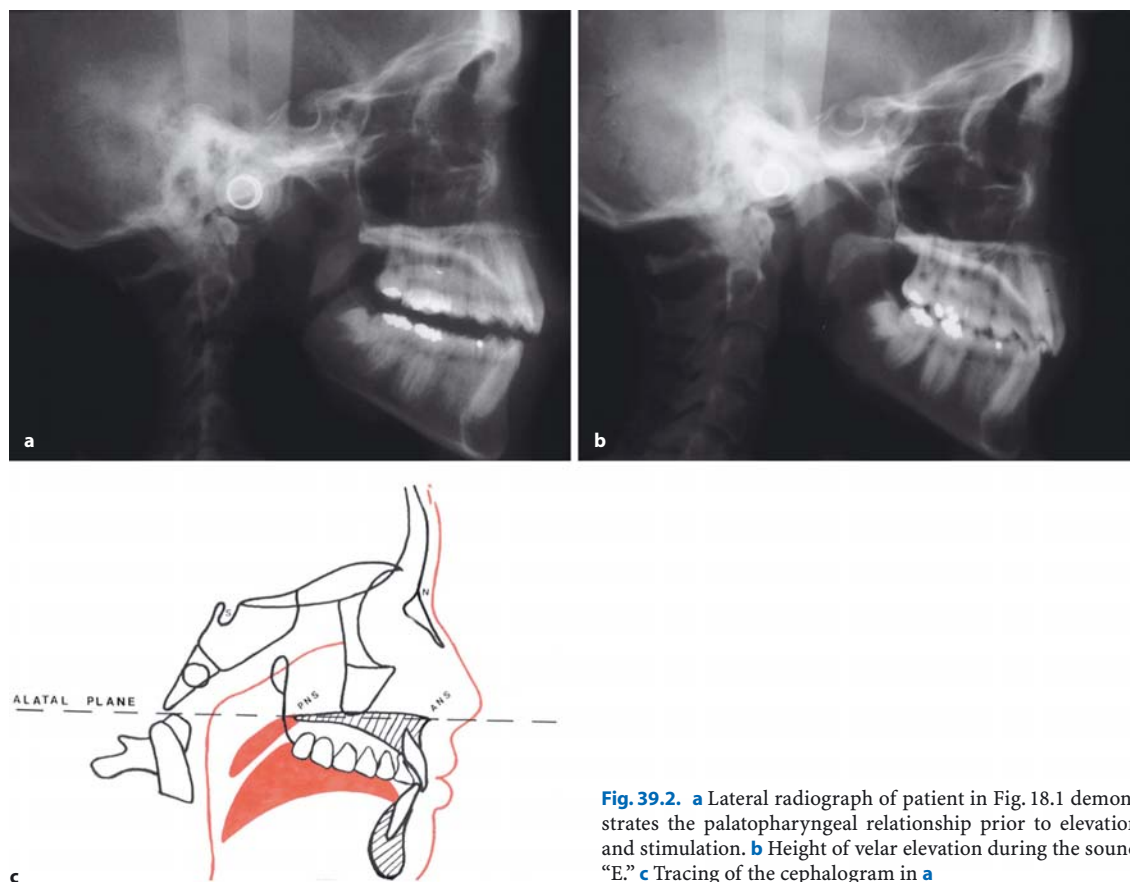


Fig. 39.2. **a** Lateral radiograph of patient in Fig. 18.1 demonstrates the palatopharyngeal relationship prior to elevation and stimulation. **b** Height of velar elevation during the sound "E." **c** Tracing of the cephalogram in **a**

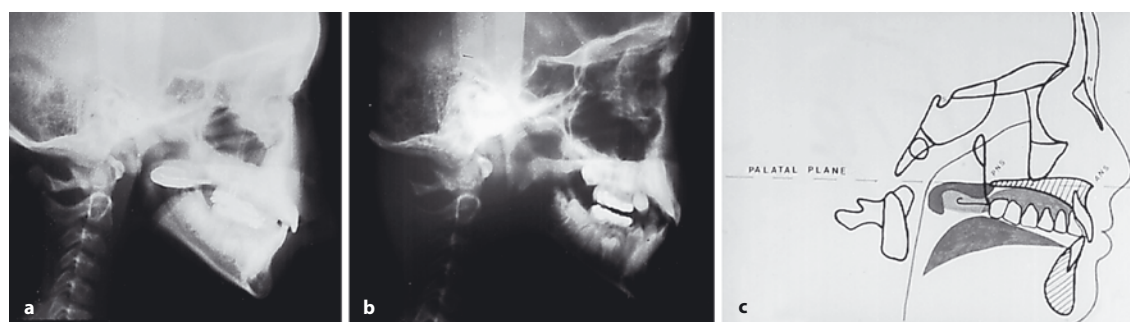


Fig. 39.3. **a** Radiographic view of the palatal lift prosthesis of patient in Fig. 18.2 in position. Note the degree of palatal elevation. **b** Increased mobility of the soft palate after 1 year of prosthetic stimulation. Pharyngeal flap surgery was done after

14 months of soft palatal stimulation, after which the lift prosthesis could be discarded. **c** Cephalometric tracing of the palatal lift prosthesis and the degree of velar elevation accomplished by the lift

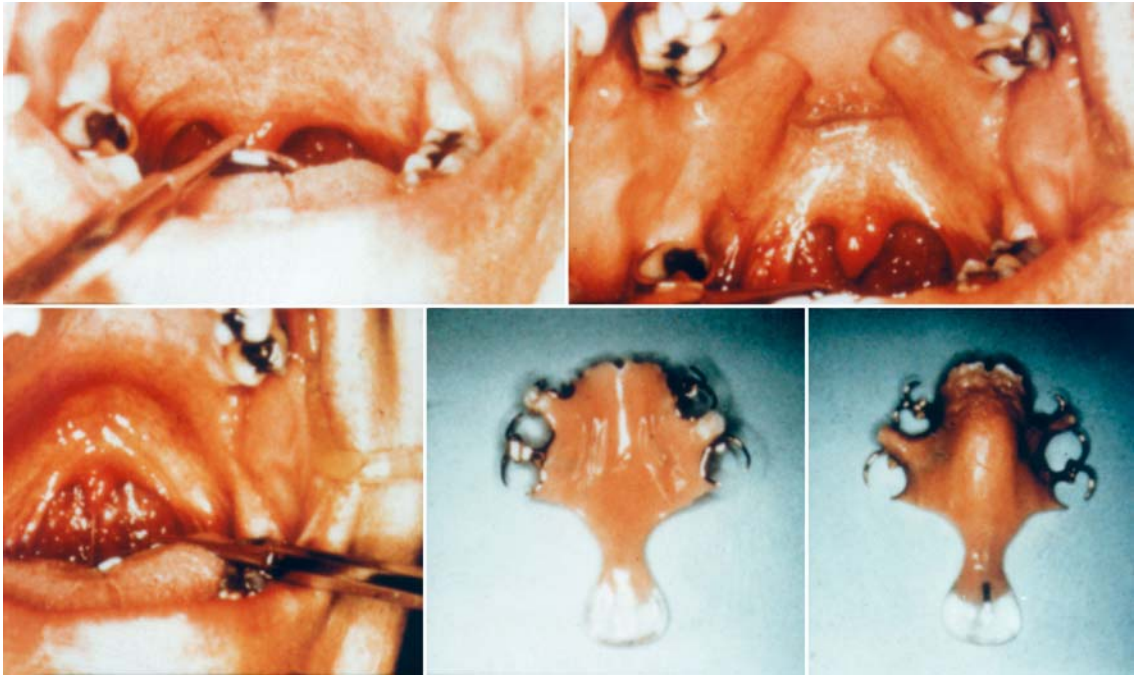


Fig. 39.4. *Top left:* Patient with palatopharyngeal incompetency in which the soft palate is paralyzed as a result of neurologic involvement after an accidental head injury. *Top right:* Palatal

lift in position. *Bottom left:* Increased soft palate elevation after 6 months of prosthetic velar stimulation. *Bottom right:* Oral and palatal view of the lift prosthesis

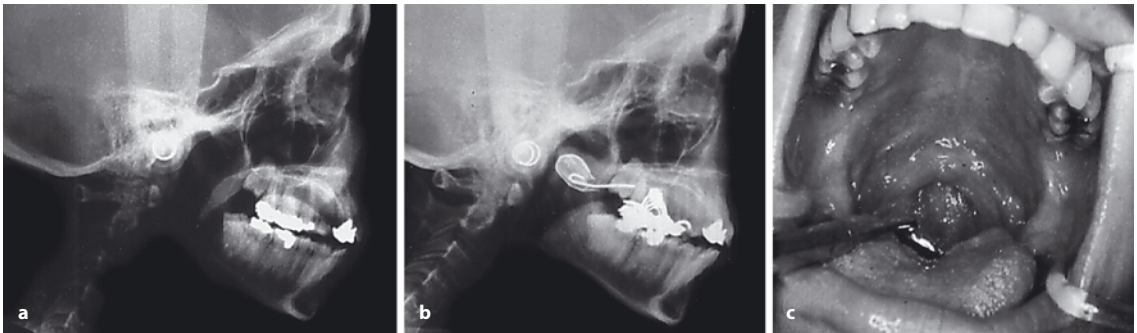


Fig. 39.5. **a** Lateral radiograph of the patient in Fig. 18.4 prior to stimulation saying “E.” **b** The palatal lift prosthesis in position elevating the soft palate. **c** Note the increase in the degree

of palatal elevation. After 11 months of stimulation and speech therapy patient is saying “E.” Note the substantial increase in the velar elevation

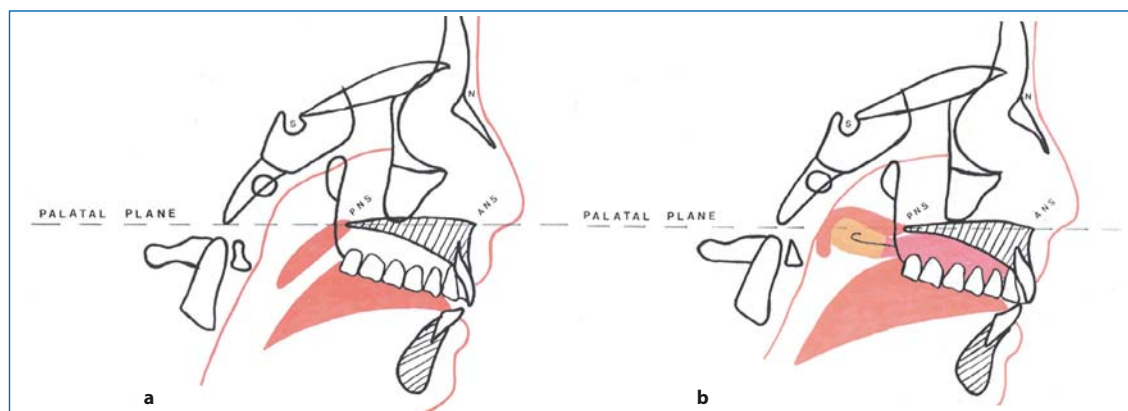


Fig. 39.6. **a** Tracing of a lateral cephalogram of the patient in Fig. 18.5 prior to soft palate stimulation by a palatal lift prosthesis. **b** Tracing of the palatal lift prosthesis and elevated soft palate

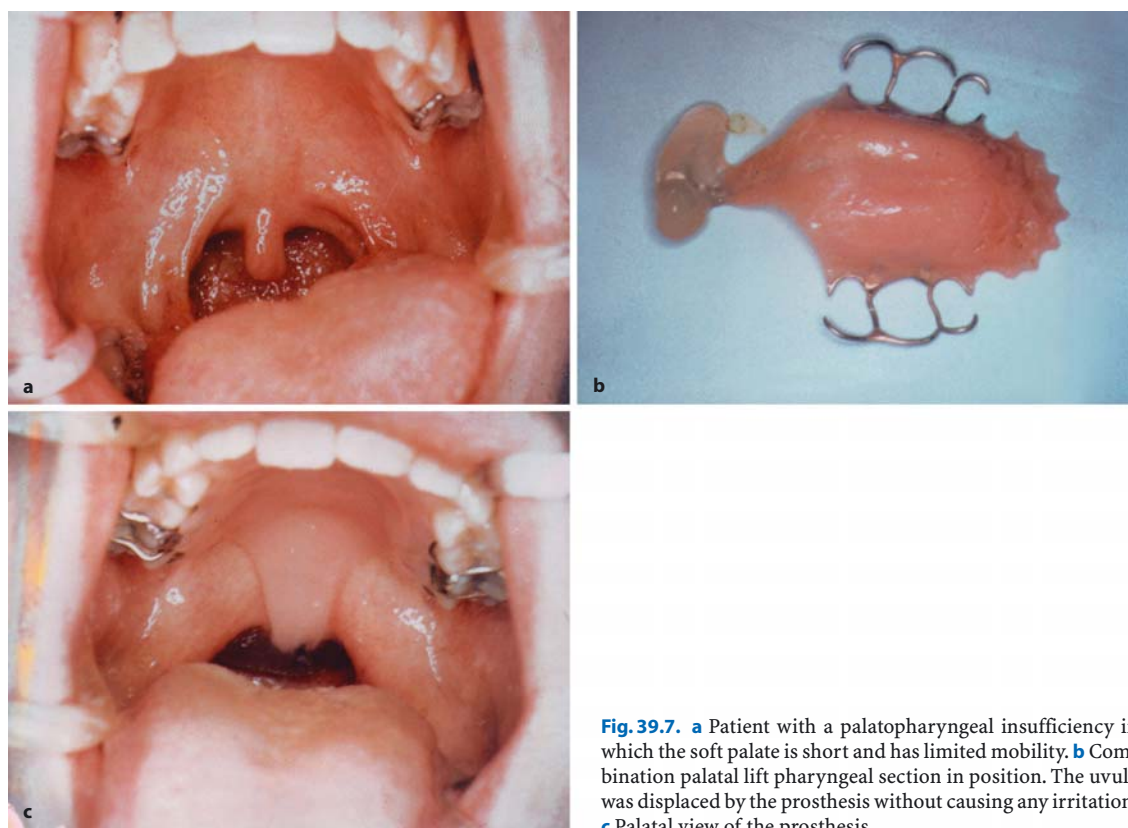


Fig. 39.7. **a** Patient with a palatopharyngeal insufficiency in which the soft palate is short and has limited mobility. **b** Combination palatal lift pharyngeal section in position. The uvula was displaced by the prosthesis without causing any irritation. **c** Palatal view of the prosthesis

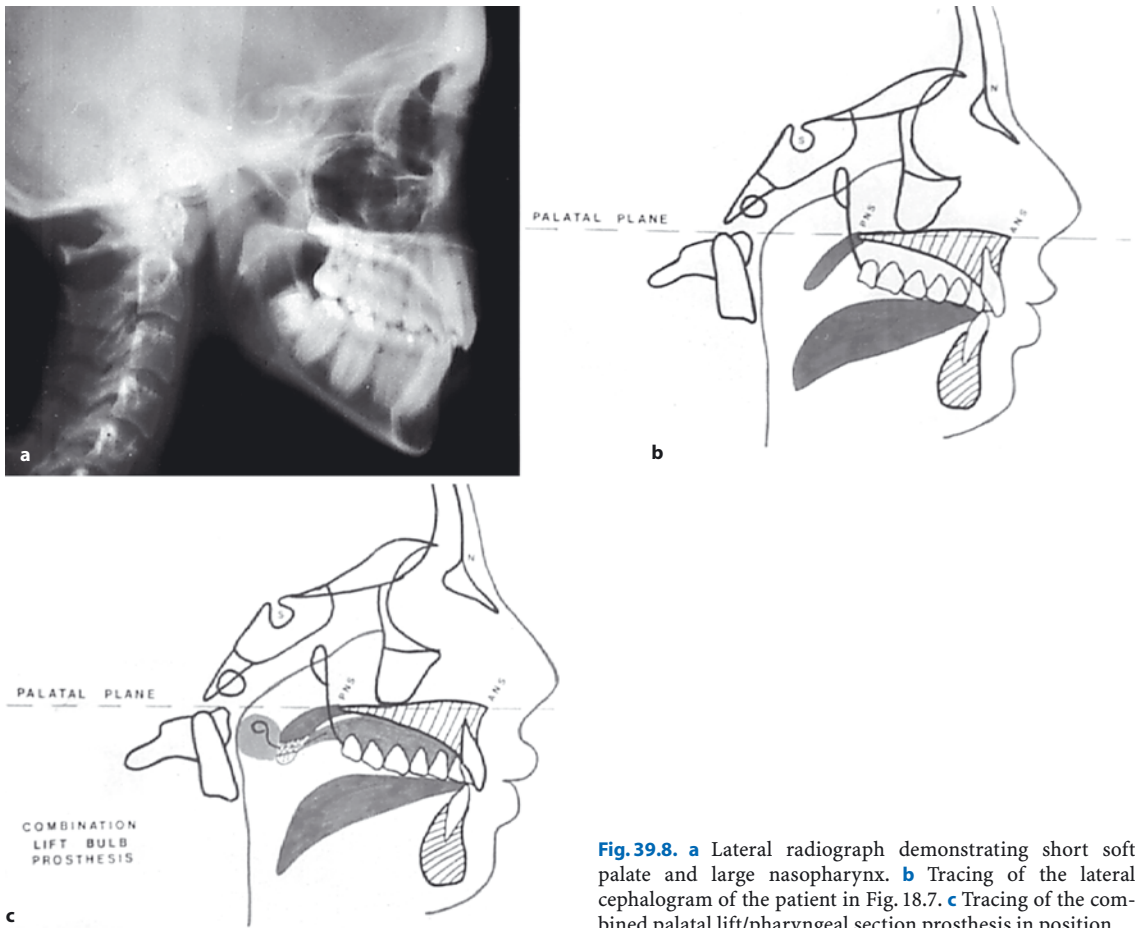


Fig. 39.8. **a** Lateral radiograph demonstrating short soft palate and large nasopharynx. **b** Tracing of the lateral cephalogram of the patient in Fig. 18.7. **c** Tracing of the combined palatal lift/pharyngeal section prosthesis in position

We have also noted variations in muscle response to mechanical stimulation. The velum of the same patient, shortly after placement of the lift, becomes more active, and after 6 months to 1 year, prosthetic stimulation and support can be discarded. Whether the increased velar elevation is the result of prosthetic stimulation or neuromusculature recovery is difficult to assess. However, we can state that, in our experience, similar patients who received speech therapy as the only mode of velopharyngeal stimulation demonstrated less functional recovery over the same period of time than patients where the prostheses were employed (see Figs. 38.6 and 38.7).

In our series of patients, we have found more marked nasal pharyngeal than velar musculature response to the prosthetic stimulation. With the velopharyngeal bulb, the patient often develops compensatory muscular constriction, requiring frequent reduction in the size of the pharyngeal bulb. In some patients, complete elimination of the bulb was accom-

plished. We could safely state that the reason for the variation in the degree of response observed in patients with velar incompetency and patients with velopharyngeal insufficiency is that we have two separate phenomena to consider. For one patient, we are trying to stimulate muscle activity by prosthetic physical therapy; for the other patient, we are attempting to create muscle build-up or constriction as a result of prosthetic placement.

39.3 Summary

1. Velar elevation should be gradual in order to put less pressure on the teeth retaining the prosthesis and to reduce the possibility of mucosal irritation.
2. Prosthetic stimulation should be initiated as soon as velar paralysis is noted, to reduce the occurrence of velar disuse atrophy.

3. The palatal lift prosthesis is used as a temporary or permanent measure for the correction of velar incompetency. As soon as adequate elevation occurs, the prosthesis is discarded. Otherwise, the patient could wear the prosthesis as a permanent supportive device.
4. Construction of the combination lift/bulb prosthesis requires a program of gradual velar elevation and molding of the pharyngeal bulb to reduce the gag reflexes and increase velopharyngeal adaptation to the prosthesis. After initial placement, modification of the velopharyngeal section becomes less troublesome to the patient.
5. Speech and myofunctional therapy should be instituted in conjunction with the prosthetic treatment.
6. Prosthetic lift and combination prostheses are more effective for patients with less severe neurological impairment and speech articulatory errors.
7. The prosthetic lift has been more effective for patients with velar incompetency without involvement of other oral pharyngeal musculatures, whereas the combination type has been more effective for patients with velopharyngeal insufficiency without marked speech articulatory disorders.

Several questions require further investigation.

1. What is the relationship between the palatal stimulation and degree of neuromuscular function and recovery?
2. What is the relationship between stimulation and degree of occurrence of disuse atrophy?
3. What is the relationship between pharyngeal stimulation and muscle constriction?
4. What is the degree of stability of velopharyngeal function and constriction after stimulation?

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40 Summary of Treatment Concepts and a New Direction for Future Palatal Growth Studies

SAMUEL BERKOWITZ

Samuel Pruzansky once said that craniofacial surgery is “an experiment on nature’s experiment.” This statement is certainly true. All facial skeletal surgery – in growing or nongrowing patients – can be regarded as an investigation of craniofacial growth, form, and function.

Because facial skeletal surgery in growing children often affects craniofacial growth as well as function, informed decisions should be made concerning which structures need to be repositioned and reformed. Based on these decisions, a treatment plan is then formulated, and a working hypothesis for successful treatment is established. Three points need to be made at this juncture. First, remembering the value of failures as learning opportunities, clinicians cannot afford to forget failures; rather they must thoroughly analyze them so they are not repeated. Second, clinical investigators must be able to explain why some surgical procedures are successful and others fail. Third, clinicians must be able to fit the proper procedure to each individual problem and be willing to work with the consequences of their choices.

Not all clefts of the lip and/or palate within the same cleft type are alike.

1. The collected serial casts and cephalometric radiographs, beginning with those of the unoperated infant and continuing through adolescence presented in this book, provide a view of the wide spectrum of variations encountered within each cleft type in its untreated state and a record of the changes that occurred thereafter resulting from natural growth or specific therapeutic procedures. Clinical experience points out one critically important, fundamental fact: All clefts cannot be lumped together as a single phenomenon. Within each type of cleft there are great individual differences in the geometry and extent of the cleft defect, and these differences are clinically significant.

In a state-of-the-art monograph in 1972, Spriestersbach and coworkers [1] wrote: “Perhaps the

greatest drawback to genetical and epidemiological research on clefts of the lip and palate has been the unfortunate tendency to lump them together.” Twenty years prior to that report, the first line in the first paper to emerge from Pruzansky’s [2] research stated: “Not all congenital clefts of the lip and palate are alike.” This statement was to become the leitmotif of his subsequent research. He took great care to demarcate samples according to varying cleft types in his designs for epidemiological, morphological, functional, and genetic research.

2. Current methods of treatment, which favor staged treatment (i.e., closing the lip at birth and the palate at a later age, in one or two stages), offer a more encouraging prognosis than those that prevailed 50 years ago.
3. The age of the patient and the type of surgery applied are two variables in determining the effect of surgery on facial growth. Quantitative and qualitative characteristics of the cleft defect, plus the general health and genotype (facial growth pattern) of the individual patient are additional determining factors. Under certain conditions, surgical repair of the palate is feasible quite early; in others, optimal conditions for repair will not become evident until a later age.
4. The natural history of children with clefts and those with specific syndromes demonstrates that some improve over time, some grow worse, and others remain unchanged despite the surgical effort.
5. Presurgical orthopedics, except for the use of a facial elastic to ventroflex the premaxilla to aid the surgeon prior to uniting the lip, have no long-term utility, and primary bone grafting has a deleterious effect on palatal and facial growth.
6. A critical review of the literature on the clinical management of cleft lip and cleft palate, together with an evaluation of the cumulative data from longitudinal palatal growth studies, has led most or-

thodontists to the following hypothesis: Conservative lip and palatal surgery facilitates rather than inhibits growth in both the maxillo-facial skeletal complex and the soft tissue of the labio-facial complex. In cleft palate cases, operative intervention which minimally involves bone growth potential will guide maxillo-facial growth in the individual in such a way that postoperative "catch-up" growth of the palate will result in acceptably normal development.

7. Within defined limits of mechanical and professional capability, the morphological and spatial relationships of the cleft palatal segments and facial growth patterns are the major determinants of the ultimate occlusion and arch form (not size). These variables, unique for each patient, could well be more indicative of the final treatment outcome than differences in the treatments employed by surgeons.
8. At the time the palatal cleft is closed, the relationship of the size and shape of the cleft space to the amount of available soft (mucoperiosteal) tissue surrounding the cleft, and the geometric relationship of the palatal processes to each other, are basic to determining the influence that scarring will have on the palatal arch form and the ability of the palate to develop normally.
9. Most skeletal malformations in cleft patients are the result of surgical procedures that have caused some growth retardation or of osteogenic deficiencies that lead to maxillary hypoplasia. All maxillary discrepancies are three-dimensional.
10. The concept that an increase in the amount of palatal scarring, beyond some critical threshold level, can reduce the palatal growth increments and cause palatal deformation appears to be valid, because the same surgical procedures, performed by the same surgeon on the same type of cleft, often lead to different palatal relationships. The reason for the different outcomes may, therefore, be due to variations in the palatal deformity at the time of surgery (i.e., the relative size of the cleft space to the size of the palatal segments that need to contribute soft tissue for cleft closure). The larger the cleft space relative to the amount of available tissue, the larger the area of denuded bone that must be left when the undermined palatal mucoperiosteum is moved medially to close the cleft space. The denuded bone heals by epithelialization, becoming a scar. The greater the scarring, the more growth retardation and palatal deformation.
11. Although the tongue has been found to occupy the cleft space and be carried high into the nose at birth, no studies have shown that abnormal tongue habits negatively affect speech development. It appears that, with the closure of the palatal cleft between 18 and 30 months, and without the use of an obturator, children usually develop good speech if the velopharyngeal closure mechanism is functionally adequate.
12. There is no documented evidence that the cleft condition interferes with body growth or that, in most instances, the palatal defect cannot be effectively treated without feeding appliances. However, obturators may be useful in some neurological disturbances when palatal closure needs to be delayed beyond 3 years of age and parents complain of feeding problems. Most pediatricians and nurses recommend the use of a soft plastic feeding bag (e.g., Playtex Nurser) or a soft plastic bottle (e.g., Mead-Johnson's nurser) with a cross-cut, normalized nipple. The use of Lamb's and Ross Laboratory nipples is strongly discouraged because of their abnormal shape and nipple length.
13. A child with a Pierre-Robin sequence should never be given an obturator, because the child's oral volume is already too small and an appliance will further compromise tongue positioning. Because the infant has a micrognathic mandible, the tongue must be carried high into the palatal cleft space during this critical early adjustment period. If an obturator or early palatal surgery is utilized for these children, it can force the tongue downward and backward, possibly closing off the airway space and interfering with breathing.
14. The use of a head bonnet with a facial elastic band or the use of elastic taped to the cheeks across the lips to reduce palatal distortion are acceptable methods to help the surgeon reduce tension at the surgical site. Such innocuous external facial forces will help bring the distorted lip and skeletal segments into a more normal relationship. This mode of treatment is acceptable to most parents and clinicians.
15. There is no proof that neonatal maxillary orthopedic appliances will stimulate palatal growth or reduce middle ear infections [3], nor has it ever been shown that these orthopedic procedures will prevent the need for future orthodontia and improve speech development. An obturator will be of some help if the cleft space remains open after 3 years of age and neurological problems interfere with feeding.
16. In many cases, protraction orthopedic forces can protrude the maxillary complex sufficiently to negate the need for surgical advancement. These forces are most efficient when applied before or during the pubertal growth spurt. After puberty, the effects change from orthopedic (bone) to orthodontic (dental) movements. The use of palatal expansion forces prior to the application of pro-

traction devices can increase the potential for orthopedic movement of the maxilla.

Once midfacial recessiveness occurs at an early age, for example after premaxillary orthopedic retraction, it will not show increased growth acceleration to spontaneously improve midfacial skeletal and dental relationships.

40.1 A New Direction for Cleft Research

Successful outcomes in the treatment of complete unilateral cleft lip and palate (CUCL/P) and complete bilateral cleft lip and palate (CBCLP) are not universally obtained, despite significant improvements in surgical techniques over the past three decades. In particular, deficient palatal growth may occur even when treatment is rendered by expert teams. The factors that contribute most significantly to unfavorable growth outcomes remain obscure.

Although the treatment of cleft lip and cleft palate has progressed markedly in the last 50 years, there is still a great need for improvement in diagnosis and treatment planning. However, to accomplish this goal our current diagnostic categories may need to be revised. The possibility that clefts that are similarly classified may react differently to the same surgical procedure must be examined. The ultimate aim of future research is to provide a better objective understanding of the reasons for, and the characteristics of, these differing outcomes, and by so doing provide a broader and more informative knowledge base for making diagnostic and treatment decisions concerning cleft lip and cleft palate.

No matter what type of treatment surgeons have favored, they have not been able to explain why their surgical method of choice, when performed on similar clefts at the same age, often yielded different results. Why some cases appear to show “catch-up-growth,” resulting in good facial and palatal form and functional dental occlusion, while others show poor facial and palatal development remains an enigma. Among the specific unanswered questions: Were the different outcomes due to different levels of skill on the part of the operators? Were there significant differences in the palatal deformity at the time of cleft closure surgery within each cleft type that should have been differentially diagnosed? And does presurgical orthopedics influence palatal growth or does it merely act to reposition palatal segments?

Catch-up-growth has been defined by Hughes [4] as growth with a velocity above the statistical limits of normality for age during a defined period of time. Such an increase in the rate of growth, before and after palatal surgery, with or without neonatal maxillary

orthopedics, may allow the palate to attain its normal adult size or, with reduced velocity, the palate may still fail to do so. The latter case is called “incomplete” catch-up growth. Wilson and Osbourn [5] showed that the duration and severity of the insult (the scarring resulting from the surgical procedure used to close the cleft space in the hard palate) may positively or negatively affect the ability of the palate to recover and undergo catch-up growth. The developmental age of the infant at the time of the insult and the nature of the insult itself (extent of denuded bone left after surgery and the resulting scarring) will affect the ability of the infant to achieve complete catch-up growth.

40.2 Clinical Research

Feinstein [6] wrote:

In the biostatistical architecture of clinical research, the first operational principle is to specify the components and choose the logic of the objective of the research. The components consist of a sequence of initial state, maneuver and subsequent state. The logic consists of suitable scientific judgment in the decisions made to demarcate the diagnostic and prognostic conditions of the initial state of the population; to identify differentiate and prognostically correlate the diverse targets of the subsequent state; and to choose maneuvers that are satisfactory in potency, comparison, multiplicity and concurrency.

In speaking of the initial and the subsequent states, emphasis will be placed on studies of casts starting at birth and extending through adolescence.

40.2.1 Initial State

The size and form of the palatal segments are measured serially starting at birth and divided into two periods. The first period ends at surgery to close the palatal cleft. The second period includes the cleft space with the changing size of the palatal segments.

Analyses of the initial state prior to palatal surgery (end of first period) suggest that, under certain conditions, surgical repair of the palate is feasible quite early; whereas in other instances, optimal conditions for repair will not be present until a later age. In our experience, a selected number of cases with very small cleft spaces underwent palatal repair at or before 1 year of age without detriment to midface and palatal growth. On the other hand, there are cases where the cleft space is too large, compared to the amount of available soft tissue, and surgery needs to be postponed to avoid creating growth-inhibiting scar tissue.

This is an example of individualized differential diagnosis and treatment planning.

40.2.2 Maneuver: Presurgical Orthopedics and Surgical Procedures Used to Close the Palatal Cleft

If we assume that qualified surgeons within a given institution or region, practicing a specific series of techniques over a given period of time represent a constant, differences in success or failure should reside in (1) the initial state (the geometric and size relationship of the palatal segments to the size and shape of the cleft space, which reflects the degree of skeletal deficiency as well as palatal segment displacement) and (2) the facial growth pattern. Of course, the sample must separate cases subjected or not subjected to presurgical maxillary orthopedics, as well as cases utilizing various cleft closure procedures, because these variables can influence the subsequent state.

Of the three components, the maneuver presented the greatest number of confounding variables. Differences between surgeons, variance in the performance by the same surgeon from day to day and over the course of several years, and differences in techniques, which are difficult to identify and compare, complicate the analysis. However, our biostatisticians believe that research objectives to test the influence of presurgical orthopedic treatment and the relationship of cleft palate space to surgical outcome can be reached. It is possible to statistically test and covary for effects due to differences between and within surgeons.

As Feinstein stated, we too believe that, within certain defined limits, the success or failure of the surgical procedure depends more on the initial state than on the variables inherent in the maneuver. To put it another way, we expect that subtle differences among patients will be more prognostic of the subsequent state than differences between surgeons.

Serial facial and palatal growth studies starting at the newborn period [7] have shown that too many factors were operating in relation to the patients under study to permit the formulation of simple, all-inclusive rules, such as any suggestion regarding the age at which clefts of the palate should be repaired. Berkowitz [7] therefore hypothesized that, at the time of palatal surgery, the ratio of the useful mucoperiosteal tissue available to the size of the cleft space determined the area of denuded bone left at the surgical site after the medial movement of palatal soft tissue. This area heals by epithelialization, which in turn becomes scar tissue. The degree of scarring could spell the difference between therapeutic success and failure, because it influences the palate's ultimate size (osseous plus soft tissue) and form.

If presurgical orthopedics enhance palatal growth and development, the cleft space in the 18- to 24-month period will be much smaller relative to the enlarged palatal segments than in cases that have not been similarly treated. This hypothesis needs to be tested using quantitative measurements. Only in this way will surgeons find reason to change their focus to include the size and form of the palate and the extent of the cleft defect, as well as the surgical-orthopedic procedures, in differential diagnosis.

Pruzansky [8] frequently stated that his most important contribution to the cleft palate literature was the conclusion that "cleft lip and the palate does not represent a single fixed entity subject to generalizations of description and classification and least of all to rigid therapeutic formulas." Although his clinical reports supported this conclusion, Pruzansky did not have a sufficient number of cases and proper cast measuring equipment to study the natural history of cleft palate growth in relationship to palatal surgery in order to individualize treatment planning. The question of the role and importance of tissue adequacy or inadequacy could not be explored until a highly accurate three-dimensional measuring tool and supporting CadCam software became available.

40.3 Palatal Embryopathology

Studies of clefts have produced conflicting interpretations regarding deficiency in mass and/or displacement of the palatal segments in space, as well as the effects of cleft surgery on palatal growth. Information relating to the complexities of embryonic facial development is fundamental to an understanding of the growth potential of the primary and secondary palate. Developmental studies [9, 10] have shown that the facial mesenchyme, which gives rise to the skeletal and connective tissues, originates from neural crest cells and undergoes extensive migration and interaction.

Coalescence of the facial processes results in the formation of the primary palate, which constitutes the initial separation between the oral and nasal cavities and eventually gives rise to portions of the upper lip and anterior maxilla. The exact mechanism of primary palate formation is not clear. However, most clefts of the primary palate appear to result from variable degrees of mesenchymal deficiency in the facial processes.

The suspected causes of clefts of the secondary palate are also varied. Slavkin [9] proposed several possible mechanisms:

1. Tongue resistance: The tongue, arched up between the shelves, delays palatal shelf movement.
2. Decreased shelf forces: Although there are no examples of mutant genes that can cause this, there are many teratogens for which this mechanism has been invoked.
3. Failure to fuse: This possible cause may be associated with delayed shelf reorientation.
4. Narrow shelves: This theory suggests that the palatal shelves can move normally enough to reach the horizontal, yet still be too narrow to reach each other. This condition could be explained by a more generalized deficiency of facial mesenchyme reaching the palatal area, making the hard palatal shelves and soft palate inherently smaller.

The causative factor has important clinical implications because it suggests that, in some unilateral clefts of the lip and palate, the size of the cleft space may be disproportionately very large and more variable in shape than in other clefts of the secondary palate. The velum in this cleft type also may be deficient in muscular tissue and predispose the child to velopharyngeal incompetency. Thus, it would be helpful to be able to identify infants with skeleto-muscular deficiencies at an early age (within the first 2 years of life) in order to customize the cleft closure procedure to enhance proper speech production as well as normal palatal growth and development. Obviously, a child with palatal tissue deficiency will have a different set of problems than a cleft palate patient with adequate palatal tissue and a cleft caused by failure of proper shelf force or failure to fuse.

40.4 The Neonatal Palatal Form in Complete Clefts of the Lip and Palate

40.4.1. The Effect of Muscle Forces

The normal palatal arch form is determined by the result of the compressive forces of the orbicularis oris–buccinator–constrictor pharyngis superioris muscle ring counteracted by the protrusive and expansive forces of the tongue. However, in the presence of clefts of the lip and palate, aberrant muscle forces cause the lip and palatal segments to be distorted in space. The lateral pull of the cleft lip musculature, coupled with the pushing forces of the tongue fitting within the cleft space, are unrestrained [11].

40.4.2 The Influence of Cleft Surgery on Palatal Form and Growth

When the cleft lip and/or soft palate are united, the cleft musculature forces are reversed, causing the laterally displaced skeletal structures to move medially into a more normal form. The increased tension of the facial musculature may vary in degree among patients and with the type of lip surgery employed. No attempt will be made to measure these forces; for the same reasons they are not measured in standard orthodontic treatment planning: it is impractical! The role of lip tension on palatal arch form, however, does deserve further investigation.

Slaughter et al. [12] first recognized the many anatomic variations within similarly classified clefts and suggested that there are great differences in the amount and quality of palatal tissue among the several cleft types and within any one type. The amount of palatal tissue relative to cleft size increases with growth, but the timing of this growth varies from one person to another. In some patients, the greatest proportional changes occur earlier than in other patients so that cleft space closure may have to be delayed to avoid growth-inhibiting scar tissue; such findings were verified by Pruzansky [13, 14] Pruzansky and Lis [15], Pruzansky and Aduss [16], Pruzansky et al. [17] Lis et al. [18], and Berkowitz [7]. Krogman et al. [19] observed postoperative catch-up growth in almost every case they studied and concluded that, by the age of 6 years, the maxillary complex is usually acceptably normal. Berkowitz et al. [20] and Mapes et al. [21] further reported that, after palate surgery, there may be a growth lag from 14 to 20 months, but subsequently the processes of orderly development may take over, and the rate of growth may even accelerate.

Berkowitz's observations (as Pruzansky and Aduss [16] did earlier) over the last 25 years have shown that, after the lip is united, the displaced palatal segments will assume various relationships to each other (some may overlap, others may butt join, and still others not touch due to premature contact of the inferior turbinate on the cleft side with the nasal septum). There seems to be a correlation of arch form, seen in the deciduous dentition, with the size and geometric relationship of the palatal segments at birth. For example, in complete unilateral clefts of the lip and palate, after the lip is united, cases with a very long noncleft palatal segment and a short cleft segment coupled with a small anterior cleft space are more likely to have the segments overlap. Other variables such as steepness of the palatal slopes and the adequacy of tissue need to be considered as well.

40.5 The Need for Three-Dimensional Measuring Techniques

Assessing the geometric form of the palate prior to closure of the cleft space will enable recommendations to be made, not only for the most beneficial surgical procedures, but also for the most opportune time to perform palatal cleft closure surgery. For example, various surgical procedures to close the palatal cleft, such as those using the V-Y and the von Langenbeck surgical techniques, involve both the anterior-posterior and/or medial movement of mucoperiosteum from the right and left palatal segments. Movement of the palatal mucoperiosteum leaves areas of denuded bone at the line of incision that heal by contraction and epithelialization (scarring). The concept that an increase in the amount of palatal scarring, beyond some critical threshold level, can reduce the palatal growth increments and cause palatal deformation would appear to have validity, because the same surgical procedure, performed by the same surgeon on the same type of cleft, but with different cleft space size, often leads to different palatal relationships. One of the reasons for the different outcomes, therefore, may be variations in the palatal deformity at the time of surgery, more specifically, the size of the cleft space relative to the size of the palatal segments that contribute soft tissue for cleft closure.

Quantitative information regarding the normal palate is noticeably sparse because of measuring limitations inherent in using various forms of calipers and rulers. Some linear two-dimensional studies on the form of the newborn arch were performed by Ashley-Montague [22], Sillman [23], Richardson [24], and Brash [25]. Their measurements, limited to maximum breadth, maximum length, maximum posterior breadth, and maximum lateral sulcus breadth, produced two-dimensional tables starting at birth.

Xerographic studies of casts were an advance over previous measuring systems, because they permitted a more accurate description of two-dimensional changes in surface area. Huddart [26, 27] concluded from these measurements that, in complete unilateral clefts of lip and/or palate (CUCL/P), the palatal surface area is deficient by age 16 compared with a normal population of the same age. Huddart suggested that presurgical orthopedics may actually hinder palatal growth. In 1971, Mazaheri et al. [28] reported on changes in arch form and dimensions associated with unilateral clefts of lip and palate and cleft palate. They found a significant pattern of anteroposterior and lateral growth retardation immediately after surgical treatment. Stockli [29], who was very critical of his own research approach, reported that there are

great limitations in the use of xerography for the study of cleft palate casts. He emphasized that arch form must be considered in the treatment of an infant with complete cleft of the lip and palate, and he recognized that three-dimensional measurements would be more appropriate for longitudinal and comparative studies.

At present in the realm of cleft lip and cleft palate therapy, treatment planning is at best an “educated art.” Clinical reports of various treatment protocols, emanating from the many and widely separated cleft lip and palate treatment centers, are usually anecdotal and understandably supportive of the clinics’ own treatment concepts. Although the protocols may differ significantly, the authors tend to be satisfied with their own patients’ facial, dental, and speech outcomes, all of which encourages few if any innovations in treatment approaches.

Certain questions inevitably arise: Do several different surgical procedures yield universally acceptable results that allow for normal palatal development? Are the outcome reports self-serving or can there indeed be a variety of effective surgical procedures? In cases of undeniable failure, what were the errors, if any, in diagnosis and treatment planning? In assessing failures, most surgeons focus solely on the surgical skills and/or surgical protocols involved, but this leaves other possible explanations unexplored. In recent years, it has been suggested that variations in the physical characteristics of the deformity – the geometric relationship of the palatal segments to each other at birth and the size of the cleft space relative to the amount of available soft tissue used to close the cleft spaces – may have an impact on treatment outcomes [20, 29]. Those authors highlighted the importance of three-dimensional measurements and urged that the arch form and the size of the cleft space at the time of surgery be taken into consideration in the treatment of infants with complete clefts of the lip and palate.

Lack of appreciation for the importance of the geometric relationships of the cleft palatal segments to each other has been the result, in great part, of the dearth of longitudinal records, such as serial palate casts and lateral cephaloradiographs, and an accurate palatal cast-measuring device for quantification and computer analysis of the palate’s changing geometric form. Just as the microscope uncovered critical differences in tissue pathology, a three-dimensional measuring instrument could reveal palatal geometric information that had heretofore gone unnoticed, and the importance of which has not been appreciated. Fortunately, such a measuring instrument and a significant number of dental casts are now available.

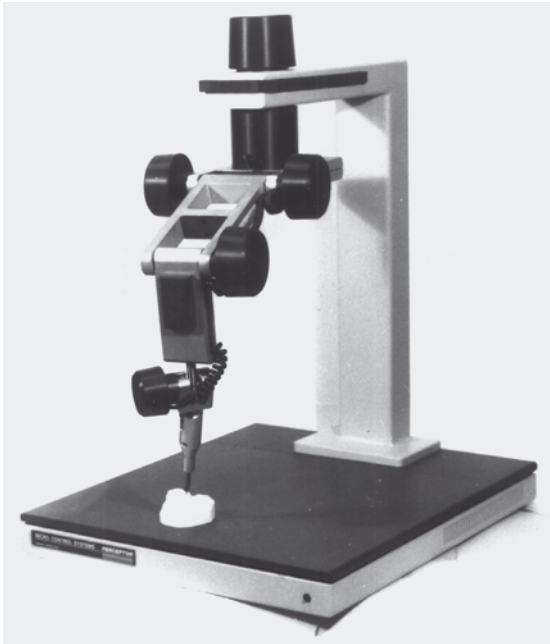


Fig. 40.1. An electromechanical digitizer used to extrapolate x , y , and z coordinates from a plaster palatal cast

40.6 Studies Using Three-Dimensional Techniques (Figs. 40.1–40.7)

Berkowitz [30] initiated a study to determine the feasibility of using stereophotogrammetry to graphically describe the changing configuration of cleft palates. Data from the study supported the clinical impressions that palatal molding action with palatal growth, which occurred at the palate's medial border, effectively diminished the width of the cleft space. A second study (Berkowitz et al. [20]) was undertaken to further improve the stereometric technology in order to permit the investigation of a larger number of casts. A profile study of nine complete unilateral cleft lip and palate casts demonstrated that the widths of the vault space varied greatly between cases. This was followed by another investigation using an "Optical Profilometer" [31] designed and built by National Aeronautics and Space Administration (NASA) for Berkowitz under a technology utilization transfer grant. This led to the use of an electromechanical digitizer as the instrument of choice for analytical studies of serial casts designed to describe the changing geometry and size of the palatal vault, and the geometrical and size relationship between the greater and lesser palatal segments in complete unilateral cleft lip (CUCLP) and palate and the lateral palatal segments

and premaxilla in complete bilateral clefts of the lip and palate (CBCLP) (Figs. 40.5, 40.6).

Serial three-dimensional palatal growth studies to date have led Berkowitz to believe that size and geometric relationship of the palatal segments relative to the size of cleft space prior to surgery, coupled with the surgical procedure utilized, may influence the palate's subsequent arch form and size (Fig. 40.7). If it does, the surgical skill or technique is not solely responsible for the different outcomes. This might explain why different surgical procedures can be equally successful and, conversely, why the same surgical procedure can cause a different result, especially if extensive scarring has been produced (Figs. 40.5, 40.6).

The following three-dimensional palatal growth studies were recently completed. These studies can be considered forerunners of multicenter efforts still to come that will reflect on the physiological attributes of the various surgical and orthopedic treatment procedures.

40.6.1 Study 1: Analysis of Longitudinal Growth of CUCLP and CBCLP

Patients from Berkowitz's longitudinal facial-palatal growth records who did not have neonatal maxillary orthopedics were the subjects.

Eleven children with unilateral clefts and 14 children with bilateral clefts were measured for palate area in mm^2 over a period of 5 years. For the unilateral cleft group, the measurements were made at approximately 6, 12, 24, 30, and 60 months. Each child in both groups was surgically treated to close the cleft area at approximately 24–36 months.

40.6.1.1 Statistical Methods

For each child, the monthly growth rate (in mm^2/month) from 6 through 24 months was estimated by linear regression. In the unilateral cleft group, the monthly growth rate after surgical intervention was estimated by the change in palate area from 36 to 60 months. This rate was estimated in the bilateral cleft group after surgical intervention over the period of 30 to 60 months. Mean growth rates before and after surgical repair were compared within each group by the paired Student's t -test. Pre- and postsurgical differences in mean growth rates between the two cleft types were compared using the two-sample Student's t -test. In addition, growth rates and the change in growth rate before and after intervention were correlated with the estimated size of the closure at surgery.

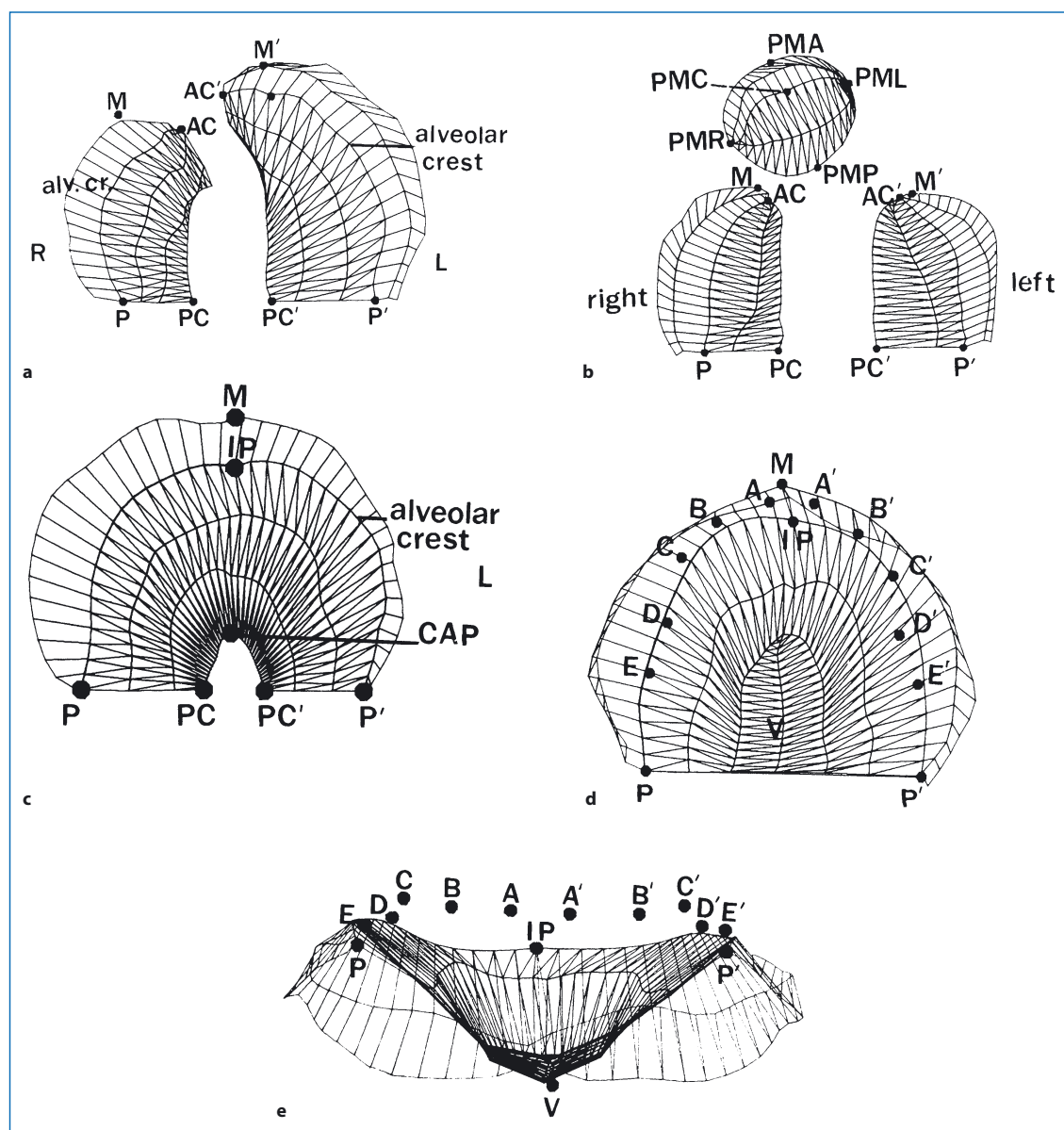


Fig. 40.2 a–e. Computer-generated images of various cleft palate types. **a** Complete unilateral cleft lip and palate. **b** Complete bilateral cleft lip and palate. **c** Isolated cleft palate. **d** Normal palate: occlusal view. **e** Normal palate: postero-anterior view. *P*, Postgingivale comparable to *PTM* [pterygomaxillary fissure on a lateral cephalograph]. It is the posterior border of the hard palate; *PC*, Landmark on the *P-P* line at the cleft; *AC*, Anterior point of the alveolar ridge at the cleft; *M*, The most anterior point of the palatal segment. *IP*, Incisal papilla point; *V*, Highest vault point; *A*, Deciduous central incisor, *B*, Decidu-

ous lateral incisor. *C*, Deciduous cuspid, *D*, Deciduous first molar; *E*, Deciduous second molar. **Palatal Surface Area.** Before cleft closure: Bounded laterally by *P* to *AC*, *P* to *Pc* and *PC9* to *P9*, *P9* to *Ac9*. After cleft closure: Includes cleft space bounded by *AC* to *AC9* and *PC* to *PC9*. In Bilateral Clefts: Anterior Cleft Space: Bounded anteriorly by the premaxilla's outer point of the alveolar crest *RPM* or *LPM* to *AC* and posteriorly by line *AC* to *AC9*. Posterior Cleft Space: Bounded by *AC* to *AC9*, *AC* to *PC*, and *PC* to *PC9*

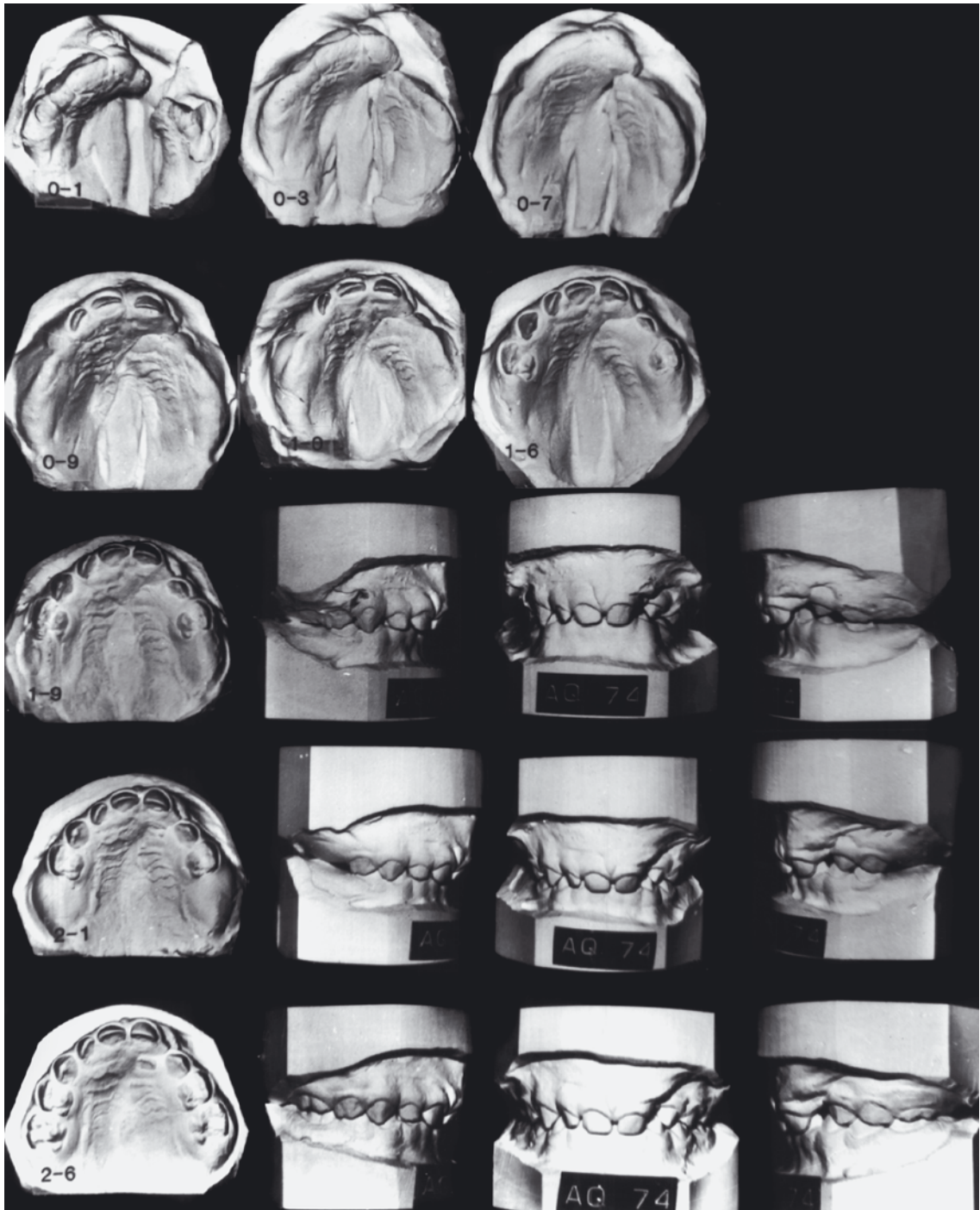


Fig. 40.3. Serial dental casts for Case JH (AQ-74) show: 0-1. Separated palatal segments soon after birth. 0-3. Palatal segments move together forming a butt joint relationship



Fig. 40.3. (continued) 0-7, 0-9, 1-6, and 1-9. What appears to be a “collapsed” state is not so. 2-1 and 2-6. The buccal teeth are in an ideal occlusal relationship. 10-0, 10-5, and 10-8. Palatal

growth maintains the excellent palatal arch relationship. The central incisors were brought together at 8 years of age prior to secondary alveolar bone grafts

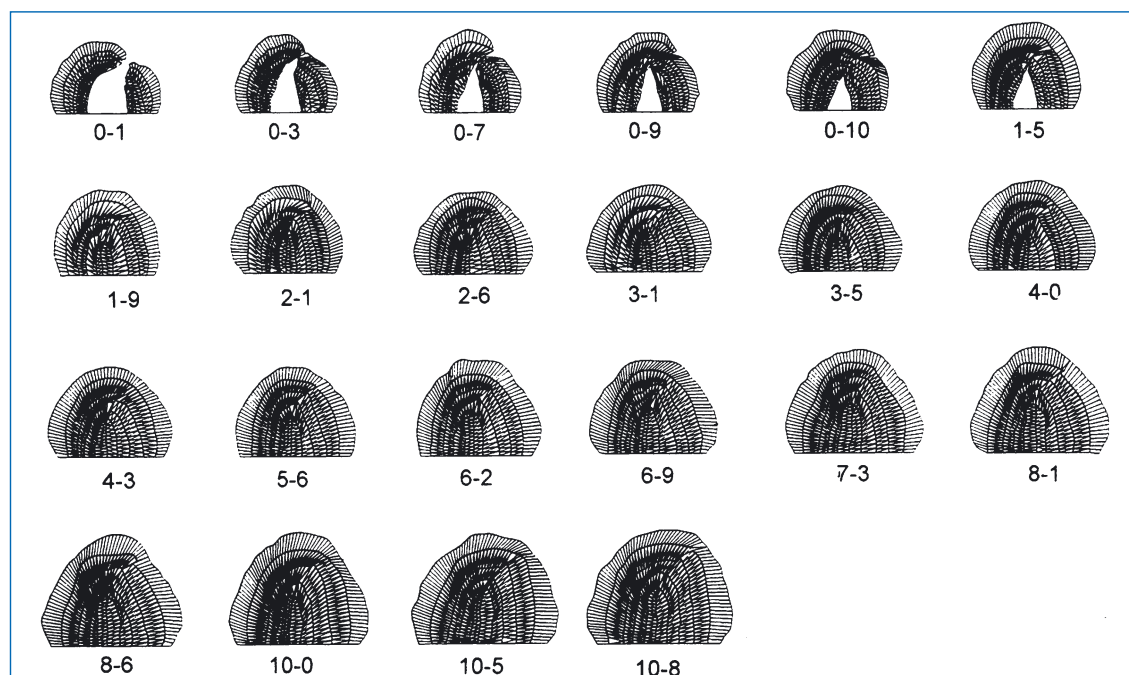


Fig. 40.4. Case JH AQ-74. Computer-generated images of serial casts drawn to scale. This series demonstrates the decrease in left spaces associated with an increase in palatal size

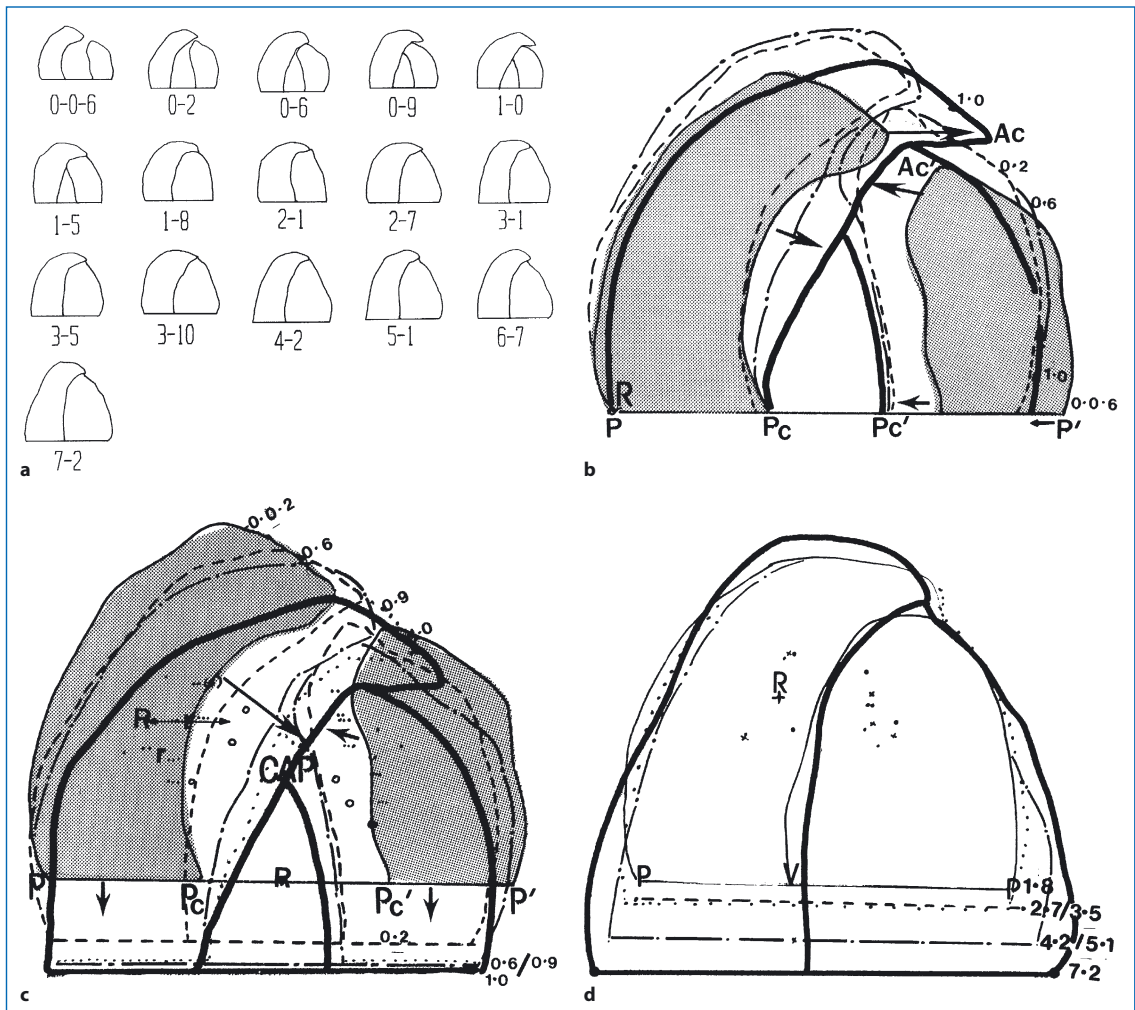


Fig. 40.5 a–d. Computer-created serial casts drawn to scale from birth to 7 years and 2 months. **b** Outline tracings at 6 days, 2 months, and 1 year of age superimposed on the baseline P-P1 and registered at midpoint of the line. This illustration shows the medial movement and changes in size of the palatal segments. **c** The same palatal segments are superimposed on the

palatal rugae to show the amount and direction of palatal growth and movement brought on by uniting the lip. From 2 days to 1 year of age. **d** Outline of palatal segments from 1 year and 8 months of age to 7 years and 2 months. This illustration shows that most of the palatal growth occurs posteriorly with a slight increase in width with little anterior bony apposition

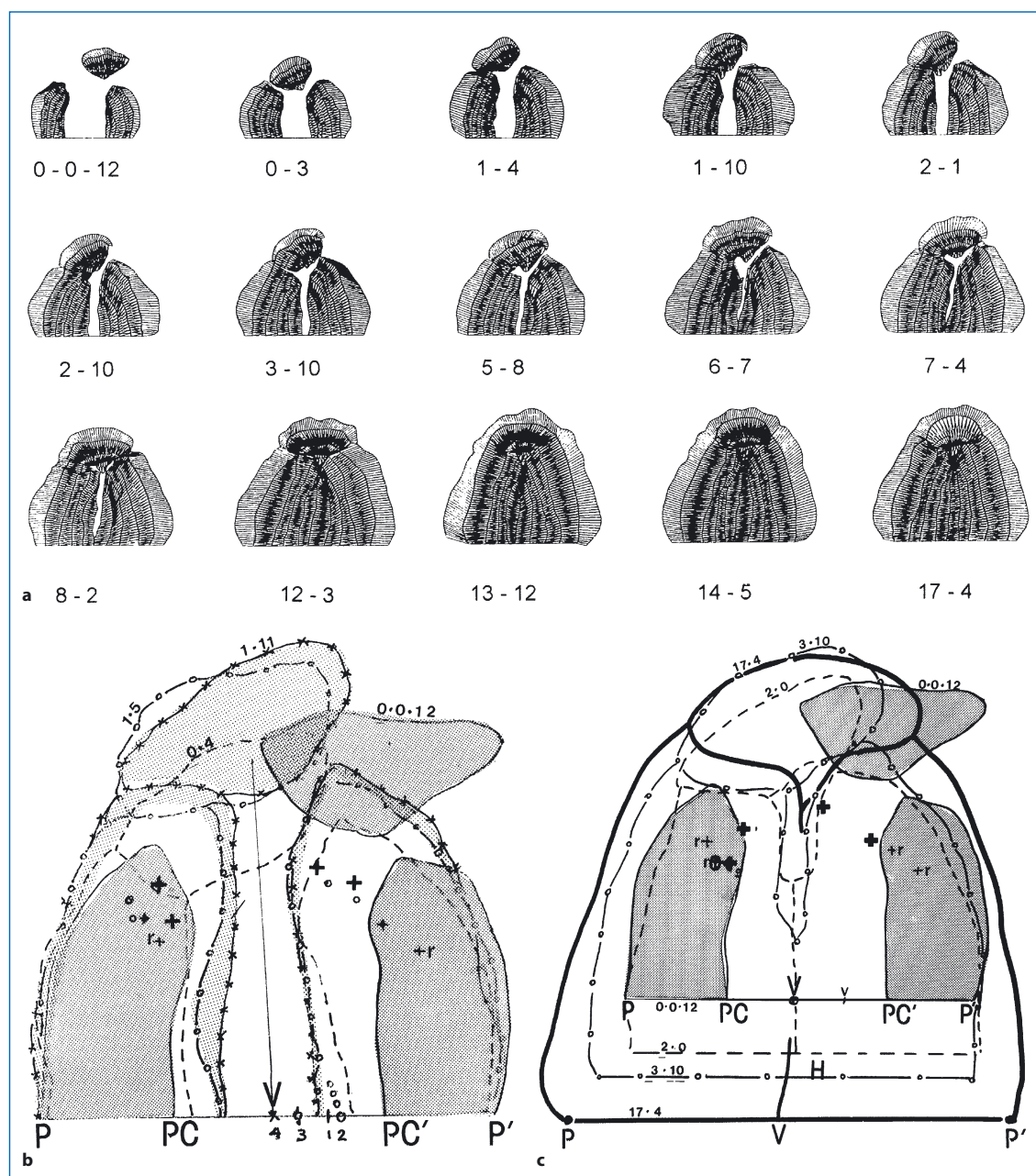


Fig. 40.6 a-d. Case PM (K-22). **a** Computer-drawn serial casts of a complete bilateral cleft lip and palate drawn to scale showing quantitative and geometric palatal changes as a result of growth and treatment. No presurgical orthopedics. **b** Serial casts superimposed on the base line P to P' , registered at V (the point where the vomer meets the baseline). This shows changes

in palatal size and palatal relationships. **c** Serial casts from 12 days to 17 years and 4 months. These casts were superimposed on the rugae points with the PP' line parallel. Most of the palatal growth occurred posteriorly, with only a slight increase in midfacial protrusion compared to the initial premaxillary protrusion. The width increased in proportion to length

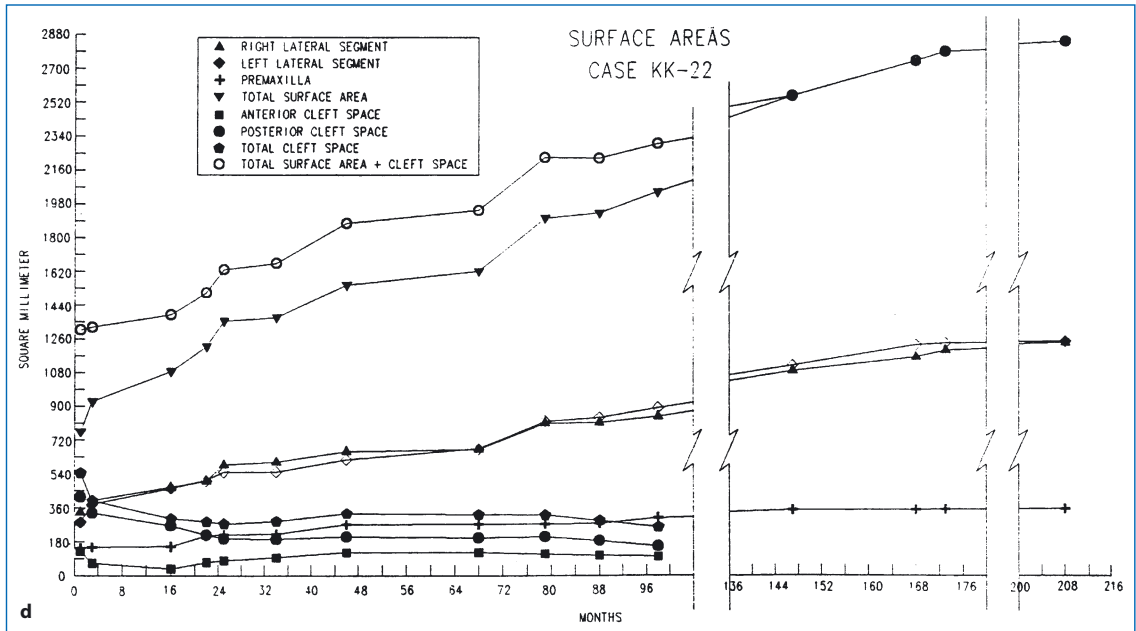


Fig. 40.6 a–d. (continued) Case PM (K-22). **d** Time sequence analysis of palatal growth in a complete bilateral cleft of the lip and palate. By 6 years, the palatal growth (including cleft space) more than doubled

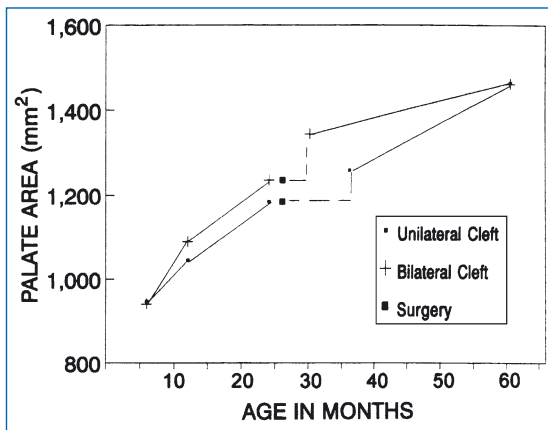


Fig. 40.7. Graph showing growth changes in surface area of 14 CBCLP and CUCLP prior to and after palatal cleft closure surgery. The palatal growth acceleration in the CBCLP was greater the first year, then similar in the two cleft types up to 25 months, just prior to palatal surgery. At the time of surgery, the cleft space in the CBCLP series was greater than in the CUCLP series. After surgery, the palatal growth acceleration curve of the CBCLP series decreased, while that of the CUCLP series increased. A modified von Langenbeck procedure to close the cleft space was used in both cleft types. The sample of cases is being increased to determine the influence of cleft size on future palatal growth and development

40.6.1.2 Results

The growth rates of the two groups before surgery (Unilateral = $12.9 \text{ mm}^2/\text{month}$, Bilateral = $15.7 \text{ mm}^2/\text{month}$) are not significantly different. However, after surgery, the growth rate of the Bilateral group ($3.9 \text{ mm}^2/\text{month}$) is significantly smaller ($p = .013$) than that of the Unilateral group ($8.7 \text{ mm}^2/\text{month}$). Comparing pre- and postsurgery growth rates, the change in growth rate for the Unilateral group (mean = 4.3 , $\text{SE} = 3.3$) was not significantly different from zero, whereas the change of growth rate in the Bilateral group (mean = 11.9 , $\text{SE} = 2.3$) was significantly different from zero.

The estimated size of the “gap” to be surgically repaired was 80.5 mm^2 in the Unilateral group and 112.5 mm^2 in the Bilateral group. These cleft sizes are not significantly different; however, this may be due to the small sample size. In a larger sample size, this difference in cleft space size probably would be significant. For all subjects combined, the remaining gap at surgery was significantly negatively correlated with the presurgical growth rate ($r = 0.52$, $p = 0.034$, one-tailed). For the Unilateral group alone, this correlation was not significant ($r = 0.31$, $p = 0.175$, one-tailed), while it was significant for the Bilateral group ($r = 0.52$, $p = 0.029$, one-tailed).

40.6.1.3 Discussion

These findings indicate that the two cleft groups had similar growth rates prior to surgery but dissimilar cleft gap sizes at the time of surgery. Following surgery, the unilateral group showed a greater, but not statistically significant, difference in growth rate. We suspect that, if the sample size is increased, the difference in growth acceleration would be significant. The bilateral cleft group showed a significantly smaller cleft space than that of the unilateral group. Both groups started out with approximately the same mean palate surface area. Before palatal surgery, both groups had similar growth rates, with the bilateral group slowing in growth after surgery. The negative correlation of the estimated cleft space at surgery with the presurgery growth rate is a measure of the validity of the measurements. To more closely analyze the patterns of growth before and after surgery, data points spaced more closely in time, especially just before and just after surgery, are planned. These data show that growth patterns can be measured and analyzed.

40.6.2 Study 2: Quantitative Study of a Patient with CBCLP Treated with Presurgical Orthopedics

This study was undertaken to graphically demonstrate the geometric changes that occurred to a palate with a CBCLP that had undergone presurgical orthopedic treatment by Kuijpers-Jagtman at Nijmegen Cleft Palate Center in The Netherlands. Besides the changes in palatal surface area and cleft space, all other dimensions can be quantified and compared with CUCLP and CBCLP cases that were treated differently at this and other institutions. A prospective clinical trial study is now underway. One of the goals of this research is to verify whether presurgical orthopedic treatment enhances palatal growth.

40.7 A New Instrument for 3D Facial and Palatal Cast Study

The Craniofacial Center at the University of Illinois College of Medicine has undertaken an extensive facial and palatal growth study using the latest technology.

Dr. Adriana Da Silveira, Director of Research, writes: The Vivid700 (Minolta) operates on a light-stripe triangulation range-finder principle. The subject's facial surface is scanned from top to bottom with a projected Class 2 laser light stripe. The position of an illuminated surface point relative to the viewpoint is obtained by triangulation. The resolution in x and y

coordinates is 200×200 range points view per scan. The reliability of this method was tested and found to be accurate. At The Craniofacial Center, 3D images are routinely captured for initial evaluation and subsequent methods. Infants are placed in the sitting position on the parent's lap. A flat background is placed between the infant and the parent. The laser surface scanner is positioned at 1 meter distant to the subject, and a series of five different scanings from different views are taken of the infant's head. Each scanning takes approximately 1 second. A custom headband is used to provide a variation in topography of the head. This allows "stitching" of the images to create a 360° three-dimensional image with the help of computer software (Polygon Editing Tools, Minolta). One facial scanning may contain 40,000 points, and a polygonal mesh is formed of all these points representing the facial surface. Cartesian coordinates (x , y , and z) from facial landmarks can be identified with the surface distance between them calculated using computer software (Measure, Minolta). These landmarks are standardized points used in physical anthropology, and their use in two and three-dimensional analysis of facial shapes is well accepted. The same software can be used to construct axial images of the head, and to measure the area and volume of the head and face or any other facial part such as the palate.

Serial palatal casts of palatal casts of cleft lip and palate subjects are scanned using the same equipment. The scanner is connected to a turntable that rotates every 60° , and scanning is performed at different views to allow for generation of a 360° composite representing the full cast. The images are stored in a personal computer and can be analyzed using the previously mentioned software programs. Surface distances, areas, and volumes can be calculated, and images can be superimposed for better visualization.

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The Eurocleft initiative consists of three elements: the Eurocleft Cohort Study, the Eurocleft Clinical Network, and the EUROCRAN Research Programme.

41.1 The Eurocleft Cohort Study

The Eurocleft Cohort Study began in the late 1980s as an intercenter comparison of the records of 9-year-

old children with complete unilateral cleft lip and palate. It sought to overcome, at least in part, some of the limitations and potential biases associated with the comparison of outcomes described in single-center reports. A full account of the methodology and findings has been presented elsewhere [1–7]. Five of the original six teams agreed to continue follow-up of the cohort until age 17. The stated surgical protocols of respective centers are shown in Table 41.1.

Table 41.1. Surgical treatment protocol of the five participating centers

	A	B	D	E	F
Birth	Presurgical orthopedics (Hotz)		Presurgical orthopedics (Extra-oral strapping)		Presurgical orthopedics (T-traction)
3 months	Lip closure (Millard, Skoog)	Lip and hard palate closure (Tennison + vomerplasty)	Lip (Varied methods and timing)	Lip and hard palate closure (Millard + vomerplasty)	
5 months					Lip closure (Modified Skoog/Tennison-Randall) and bone grafting
9 months	Soft palate closure (von Langenbeck, Perko, Wardill, Kriens)		Hard and soft palate closure (Varied methods and timing)		
18 months					
24 months		Soft palate closure (Wardill pushback)			
8–11 years	Bone grafting and hard palate closure	Bone grafting	Bone grafting	Bone grafting	

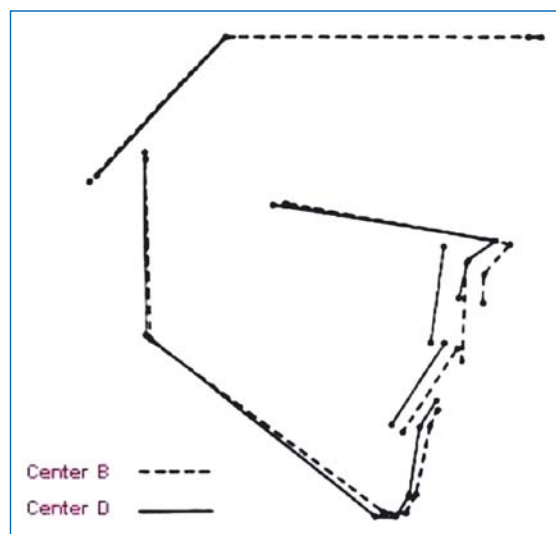


Fig. 41.1. Superimposition of mean plots for skeletal structures at age 9

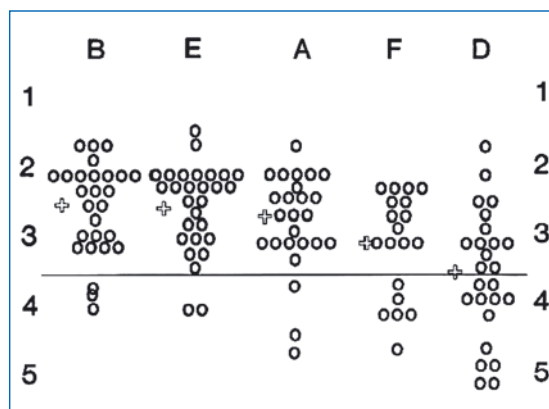


Fig. 41.2. Goslon individual patient scores at age 9 by center. A Goslon score of 1 represents excellent maxillary prominence, and a score of 5 severe maxillary retrusion. One way to consider this outcome variable is the likely future need for subsequent maxillary osteotomy; cases falling below 3.5 at this age are likely candidates for osteotomy in the late teens

41.1.1 Outcomes at Age 9

At age 9, several differences between the centers were apparent. In the main, these reflected dissimilarities of centers D and F with the remainder. Patients from center D were characterized by a flattening of the nose, reduced nasal prominence, a short upper lip, and a concave profile. Center F patients were also characterized by a retruded upper lip and relative reduction in upper face height. In center D's patients, the skeletal profile was also retrusive compared with center B (Fig. 41.1). Major differences were evident for dental arch relationship (Fig. 41.2). Whereas only 7% of cases for center E were considered to have a likely future need for osteotomy, almost half (48%) did so for center D.

It was not possible to ascribe success or failure to particular details of the surgical protocols, but poor outcomes appeared to be related to decentralized services without consistent protocols.

41.1.2 Follow-up

The aims of the follow-up were: to quantify the burden of care imposed by respective protocols; to see whether the ranking of centers for different outcomes at age 9 was predictive for equivalent outcomes at age 17; to assess patient/parent satisfaction with care, and to explore interrelationships with outcome and burden [8–11]. A separate comparison of speech outcomes was carried out at age 11–14 [12].

41.1.3 Survey of Treatment Experience

The amount of treatment provided by the five different teams in 1976–79 was remarkably different (Table 41.2). Most notable was the lengthy hospital stay associated with presurgical orthopedics at that time in centers D and F. The subjects in center D also had more orthodontic visits for treatment and review, and for the overall number of surgeries compared to the other centers. From discussion with these centers it would seem that the reason for the large differences in the intensity of treatment was not primarily related to clinical need, but rather differing beliefs and historical practices that had shaped the clinical protocols of the period.

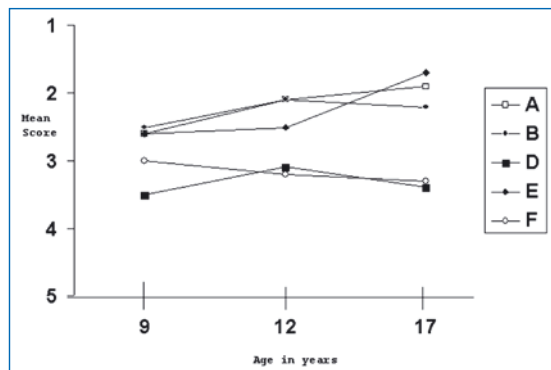
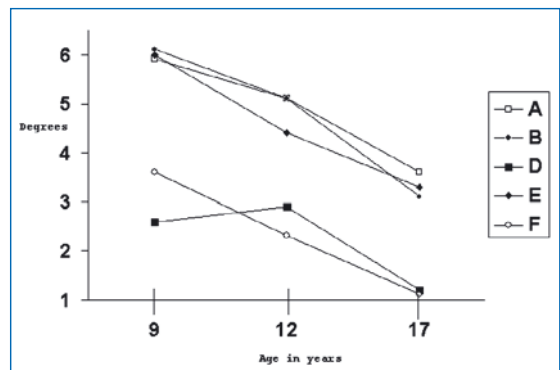
41.1.4 Consistency of Outcomes Over Time

The statistical analysis used to compare the five centers was a general linear mixed model applied to longitudinal data [13]. Variance terms were included in the model to account for between-subject variation in the intercept as well as fixed factor for assessment point (9, 12, 17 years) and center. Full details are reported elsewhere [14].

As Figure 41.3 indicates, the scores for dental arch relationship tended to improve in centers A, B, and E, but not in D and F. There was a consistent relationship over time for most cephalometric variables, e.g., soft tissue profile (Fig. 41.4) and for nasolabial appearance.

Table 41.2. Amount of surgery, presurgical orthopedics, and orthodontic treatment at the five centers

	A	B	D	E	F
SURGERY					
Mean number of surgeries	4.8	3.3	6.0	4.4	3.5
Mean days in hospital	33	31	60	24	26
EARLY ORTHOPEDICS					
Months of treatment	13	0	15	0	5
Number of visits	11	0	8	0	17
Days in hospital	0	0	60	0	146
ORTHODONTIC TREATMENT					
Treatment length (years)	5.6	3.3	8.5	3.5	4.0
Number of visits Treatment	52	41	54	33	47
Follow-up	11	23	42	16	25
Total	63	64	94	49	72

**Fig. 41.3.** Mean dental arch relationship scores at ages 9, 12, and 17 years for participating centers**Fig. 41.4.** Mean soft tissue profile (angle SSS-NS-SMS) at ages 9, 12, and 17 years for participating centers

41.1.5 Lack of Association Between Outcome and the Amount of Treatment

Not surprisingly, follow-up of these five cohorts of patients from age 9 to age 17 confirmed the main finding of the first report, with some centers continuing to achieve considerably better outcome than others, at all age points. Perhaps more surprising is the lack of association between amount of treatment and final outcome (Tables 41.3–41.5). Especially ironic is the finding that the two centers with the highest intensity of early treatment (hospitalization in order to perform presurgical orthopedics) achieved the lowest rankings for eventual outcome (Table 41.3). Patients in the center with the least favorable outcomes (center

D) also experienced the longest orthodontic treatment duration and the highest number of orthodontic visits. It appears that this was partly due to the complexity of center D's orthodontic treatment protocols with almost continuous treatment from the eruption of the primary dentition, and partly to the unfavorable dentofacial outcomes of primary surgery. (It is now almost 30 years since treatment began for these cohorts of patients, and hospitalization for orthopedics has long been discontinued).

This lack of association between treatment outcome and intensity may represent a key lesson for the development of future protocols. It justifies an emphasis on simplicity, economy, and minimized burden for the patient, rather than adherence to demanding protocols with unsubstantiated promise.

Table 41.3. Relationship between outcome assessment (dental arch relationship using the 17-year-old yardstick) and amount of infant orthopedic treatment in the different centers

Objective Ranking	Center	Months of treatment	No. of visits	Days in hospital
Best	E	0	0	0
	A	13	11	0
	B	0	0	0
	F	5	17	146
Worst	D	15	8	60

Table 41.4. Relationship between outcome assessment (dental arch relationship using the 17-year-old yardstick) and amount of orthodontic treatment in the different centers

Objective ranking	Center	Treatment length (years)	No. of visits	
			Treatment	Check-up
Best	E	3.5	33	16
	A	5.6	52	11
	B	3.3	41	23
	F	4.0	47	25
Worst	(D)	8.5	54	42

Table 41.5. Relationship between outcome assessment (dental arch relationship using the 17-year-old yardstick) and the mean number of surgeries per patient in the different centers

Objective ranking	Center	No. of surgeries
Best	E	4.4
	A	4.8
	B	3.8
	F	3.5
Worst	D	6.0

41.1.6 Lack of Association Between Outcome and Satisfaction

Perhaps the most perplexing finding of the series is the inconsistency between objectively rated outcomes and patient/parent satisfaction. We observed instances where the highest levels of dissatisfaction with treatment outcome were reported by subjects attending the centers with the best objective ratings (Table 41.6). The possible reasons for this disparity have been discussed elsewhere [9] and highlight the need for concerted work on the understanding and measurement of patient satisfaction and the provision of more holistic models of cleft care.

41.1.7 The Benefits of Intercenter Comparison

It goes without saying that professionals entrusted with the provision of health care have an obligation to review the success of their practices and where shortcomings are revealed, to take remedial action. Such efforts should constitute a continuous cycle, sometimes known as clinical audit. This has been defined as “the systematic critical analysis of the quality of care including procedures for diagnosis and treatment, the use of resources and the resulting outcome and quality of life for the patient” [15]. Often, efforts in clinical audit are divided into evaluating the process of care (the way in which it is delivered) and the outcomes of care (what is achieved). Cycles of outcome audit are more easily established when the intervention is common and the consequences are clear-cut and quickly observable. Cleft audit therefore involves a considerable challenge because of the lengthy follow-up required, the complexity, subtlety, and number of relevant outcomes, and above all, the relatively low number of cases.

However, intercenter collaboration still offers significant advantages, by providing insights into the processes and outcomes of treatment of comparable services elsewhere, the establishment of future goals, and the exchange of evidently successful practices.

Table 41.6. Relationship between objective ranking of nasolabial outcome and patient dissatisfaction

Objective ranking		Percentage of respondents dissatisfied with nasal appearance		Objective ranking		Percentage of respondents dissatisfied with lip appearance	
Best	↑	A	64	Best	↑	B	14
		E	32			A	41
		B	14			F	6
		D	45			E	42
Worst	↓	F	33	Worst	↓	D	16

The participation of two UK centers in the 9-year-old comparisons in this series was an important stimulus for the UK's national assessment and reorganization of cleft services that subsequently took place [16].

Perhaps the greatest benefit of intercenter comparisons is the co-operative spirit they foster and the gradual diminution of rivalry that occurs. It is no coincidence that the majority of the participants of this cohort study and the related Scandcleft study [17, 18] are now participants in the same multicenter randomized trials. Working together also allows the sharing of past successes and failures and specifically facilitates fruitful joint working, such as the development of rating scales and the formulation of new research questions.

41.1.8 Audit vs. Research

A fundamental limitation of intercenter comparisons is that they cannot distinguish between the influence of different individual elements of a center's protocol on its outcomes or between its protocols and the influence of the personnel who deliver that protocol. In the present series, both centers with the poorer outcomes employed presurgical orthopedics (of different kinds), but their disappointing results might have been due to many other reasons such as the role of low-volume surgeons in center D and to the use of early bone grafting in center E, rather than to an adverse effect of early orthopedics. What it does tell these centers is that elsewhere, better results are being achieved without orthopedics and the additional burden and cost that this imposes. (In any event, both centers changed in several ways; D discontinued early orthopedics, adopted the protocols of center E, and changed the way care was delivered from low- to high-volume specialists, and center F discontinued early bone grafting as well as early orthopedics).

While a series of intercenter comparisons with large numbers of cases would eventually allow one or another center to emerge as the highest achiever for particular outcomes, this would be of limited value to

the clinical community as a whole. Only protocols can be transferred, not clinicians. Inevitably, the definition of good or bad protocols, or good or bad elements of protocols requires the explicit arrangements of a randomized trial. And this is the main distinction between audit and research: clinical research (principally clinical trials) determines optimal procedures that can be transferred to an infinite number of centers; clinical audit determines whether they have been transferred successfully.

41.1.9 Alternatives to Intercenter Comparisons for Routine Clinical Audit

Despite the advantages of collaborative work listed above, there are two important limitations to their use as the routine method of clinical audit. Firstly, multiple between-group comparisons increase the sample of cases required and secondly, the logistic challenges and costs may be an issue. For most teams, reassurance that they are achieving outcomes in the mainstream of competent practice may be sufficient, rather than knowing their relative ranking against a selection of other centers. Several possibilities may be considered.

41.1.9.1 Good Practice Archive

One strategy would be to assemble an archive of relevant clinical records that are considered to be representative of good practice, perhaps drawn from the consecutive cases of respected centers. Provided other centers collect equivalent consecutive records, matching of cases on relevant characteristics would enable comparisons to be made. This could be performed by the center in question 'behind closed doors' or, if preferred, with more transparency. For example, the center's cases could be mixed with the archive cases and independently rated, or rated by a blinded panel involving the center's own personnel. With appropriate

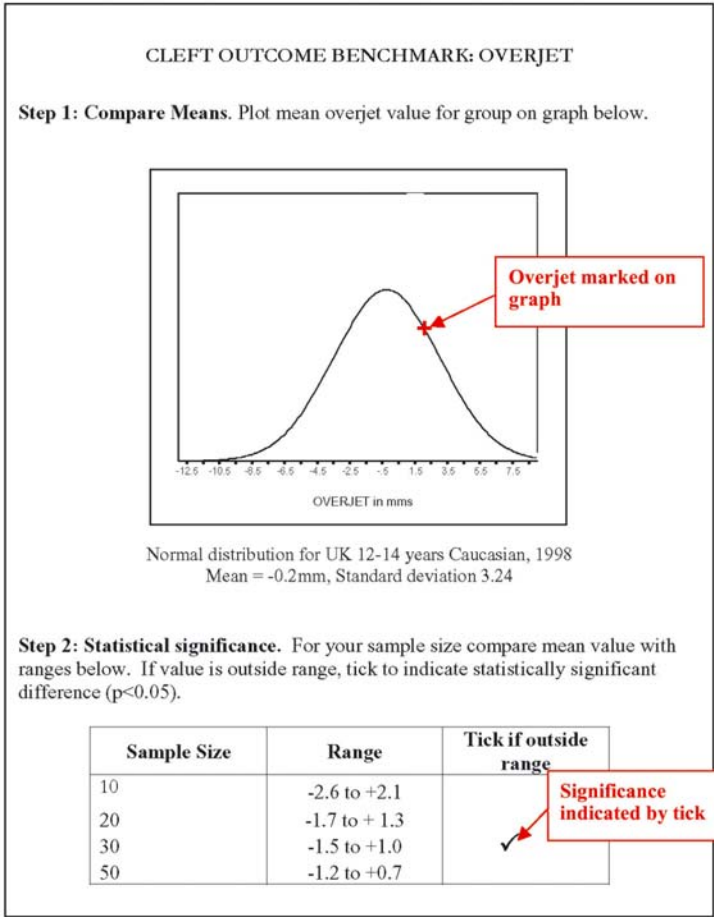


Fig.41.5. A simple two-step system to check outcome for a series of consecutive patients with UCLP based on the mean value for overjet on the noncleft central incisor. No special equipment or statistical skills are required. Any appropriate distribution may be selected as the reference base. From [20] with kind permission

technology, such exercises could be managed via the Internet to maximize efficient use of time. However this must be set against loss of the benefit of face-to-face professional interaction and discussion. (See Sect. 41.3 EUROCRAN below).

41.1.9.2 Registries

An alternative or complementary approach is the use of a registry. Prospective entry of newborn patients would have the particular advantage of establishing a list of consecutive cases that could be used to affirm that follow-up and exclusion bias are not confounders of later comparisons.

41.1.9.3 Benchmarks

The simplest system would be for teams to obtain a minimal set of objective measurements on their cases that are subject to negligible measurement error, e.g., incisal overjet [19]. These could then be compared

with tables or graphs summarizing 'good practice' outcomes. One way of doing this would be to convert values for the reference data to a normal distribution curve. Individual centers could then plot their mean value for a particular characteristic on the curve to determine the extent of any difference from the norm and its statistical significance [20](Fig. 41.5).

41.1.10 International Agreement on Record Collection and Comparison Methodology

All of the above would be facilitated by a broad consensus approach. At a recent meeting held under the auspices of the World Health Organization [21] a global consensus on recommendations for record keeping was agreed (www.who.int/ncd/hgn/publications.htm). This defines *minimum* record keeping across a range of cleft types and treatment episodes for centers that might wish to participate in future international comparisons.

In the meantime, urgent work is needed by cleft researchers to establish norms for outcomes over the range of cleft care and to undertake collaborative work to refine comparison methodology. Further work on the long-term reliability of early outcome assessment is also a high priority if the elimination of unsuccessful protocols is to take place more rapidly. Longitudinal archives from cleft clinics from around the world could make a significant contribution in this work, by defining which early measurements are most likely to be predictive over time.

41.2 Eurocleft Clinical Network

In the 1990s, competitive funding initiatives within the European Union to encourage multistate cooperation were announced. A successful application by the participants in the cohort study provided an opportunity to broaden its original goals. The project ran for three years between 1996 and 1999 within the member states of the EU. Teams from Central and Eastern European states joined the network for a further year [22].

The ultimate goal of the project was to improve the effectiveness and efficiency of care for European children with clefts.

41.2.1 Creation of the Network

Each member of the project Steering Group had responsibility for linking with cleft professionals from a specific group of countries where they had particular expertise in terms of language or experience. This strategy facilitated a nomination process among the registered teams who were asked to suggest individuals from each of their countries to act as a key delegate and provide a general link to the network. The criteria for nomination included the requirement that the individual should be an expert in the field of cleft care with a good knowledge and general overview of cleft care organization in their country. National representatives were chosen on the basis of nomination by their national colleagues and a spread of clinical specialty in order to secure a multidisciplinary breadth to the network.

41.2.2 Development of Consensus Recommendations

It was evident from previous investigations that the quality of cleft care would vary enormously between one region and another arising from organizational or surgical factors. It was considered inappropriate to

prescribe specific technical protocols on the timing, sequence, and design of surgical procedures, orthodontic treatment, speech therapy and so forth as many of these choices remain highly controversial and practices currently vary to a great extent across Europe.

However, it was hoped that quality improvement of an organizational nature could be promoted by recommendations regarding policies and practice guidelines. Therefore, the Steering Group and the national representatives forming the Eurocleft network came together in a series of workshops to identify areas of agreement and to draw up minimum recommendations in the form of Policy Statements [22] (www.eurocran.org/content.asp?contentID=776) and Practice Guidelines (www.eurocran.org/content.asp?contentID=777&sid=142954).

It was proposed that quality improvements in surgical skills would best be achieved by the promotion of clinical audit. If the methodology for assessing outcomes could be designed and established by consensus, this would allow European centers to evaluate their own quality of care, compare it with that achieved by other centers, and initiate local quality improvement. Relevant guidelines were agreed at further workshops and circulated to all registered centers of the project, national health authorities and governments [22] (www.eurocran.org/content.asp?contentID=779&sid=143236).

41.2.3 Survey of European Services

A European register of cleft centers was assembled including a total of 201 centers from 30 European countries. A survey across the network revealed great diversity in national policies, service organization, and clinical practices.

41.2.3.1 Models of Care and National Policies

The greatest challenge to the delivery of adequate services across much of Europe appeared to be organizational. Compliance with key recommendations such as the creation of fully comprehensive teams with a sufficiently large caseload to optimize clinical experience and outcome evaluation (e.g., 40 new cases per year per surgeon, orthodontist, and speech therapist) is being achieved in a minority of countries. From the workshops it was clear that the greatest obstacles to progress were human, not economic.

In some countries, policies concerning the provision of cleft care are well established. In Denmark, the provision of primary surgery by one surgical team has been formalized in law since 1937. More recently, in

Table 41.7. Sequence of operations for the repair of unilateral cleft lip and palate

First operation	Second operation	Third operation	Fourth operation	%
Lip closure	Hard and soft palate closure			42.8
Lip closure	Soft palate closure	Hard palate closure		15.3
Lip and hard palate closure	Soft palate closure			10.4
Lip and soft palate closure	Hard palate closure			10.0
Lip, hard and soft palate closure				5.0
Lip closure	Soft palate closure	Hard palate closure and alveolar bone grafting		3.5
Lip and soft palate closure	Hard palate closure and gingivo-alveoloplasty			2.5
Lip and alveolar closure	Hard and soft palate closure			2.0
Soft palate closure	Lip and hard palate closure			2.0
Lip adhesion	Lip closure	Soft palate closure	Hard palate closure	1.5
Lip and alveolar closure	Soft palate closure	Hard palate closure		1.0
Lip adhesion	Lip, hard and soft palate closure			1.0
Lip adhesion	Lip and hard palate closure	Soft palate closure		1.0
Hard and soft palate closure and alveoloplasty	Lip closure			0.5
Lip and soft palate	Hard palate closure and alveolar bone grafting			0.5
Lip adhesion	Lip closure	Hard and soft palate closure		0.5
Lip closure	Soft palate closure	Gingivo-alveoloplasty	Hard palate closure	0.5
Total				100

Norway, the long-standing arrangement of two national centers was confirmed by a national policy stating that no care can be provided elsewhere unless requested by one of these cleft teams. In the Czech Republic, Latvia, Slovenia, and Slovak Republic, well-established regional centers are continuing to provide comprehensive services following recent political and economic changes. In some circumstances, a move towards privatization of healthcare is considered to present the risk of possible destabilization of established centers. At the other extreme, the concept of team care is still to be adopted. National representatives from Bulgaria, Greece, Italy, Portugal, Romania, Spain, and Ukraine, for instance, reported that much care was still provided by individual clinicians, working in isolation, and not in government-funded centers.

41.2.3.2 Clinical Practices

Involvement of a wide variety of surgical specialties in cleft surgery was reported. Those mentioned most often were plastic surgery, 94 (46.7%); maxillofacial surgery, 59 (29.4%); pediatric surgery, 22 (10.9%); and ENT surgery, 4 (2%).

The clear lack of a sound evidence base for selecting treatment protocols was reflected by a remarkable diversity of practices across Europe for the surgical care of just one cleft sub-type, unilateral complete cleft of lip, alveolus, and palate. Seventeen possible sequences of operation to close the cleft are practiced (Table 41.7). Though 86 (42.8%) of teams closed the lip at the first operation and the hard and soft palate together at the second, almost every other conceivable sequence appears to be practiced somewhere. Thus the total number of operations taken to complete the closure of the cleft varied from one, 10 (5%); to two,

144 (71.1%); three, 43 (21.9%); and four, 4 (2%). Around half the registered teams employ presurgical orthopedics, of which 67 (65%) use it routinely. Mostly passive plates were used by 74 (70%) of teams and some teams, also used the plate as a feeding plate.

Not one single European team practiced the same protocol as any other (except for those participating in the clinical trial described below). That is to say, 201 teams were practicing 194 different protocols for one cleft subtype.

41.2.3.3 Consumer Associations in Europe

Of the 28 European countries involved in the network, 18 have some form of organization for parents. Such groups vary in the degree of their activity, effectiveness, and influence and take different forms. Some are functioning as officially established national associations while others are operating on a more informal basis. In Germany, for instance, there are several small regional organizations and one major group, which takes part in the annual meetings with the professional association. Denmark has a well-established group for parents which has been functioning for over 25 years. Interestingly, in Bulgaria, the parent group is working to bring together national cleft specialists with a view to instigating an association for professionals.

41.2.4 The Challenge of Improving Quality of Services

Achieving optimal standards of cleft care across Europe remains an outstanding challenge. The basic principles of care described in the Eurocleft Policy Statements and Practice Guidelines are hardly revolutionary; indeed they describe what most individuals (including those in clinical practice or government) would wish for their own children, regardless of country of birth. The potential strength of a European network is that examples of good practice can be more easily shared, challenges of a general nature can be more easily recognized and understood, and solutions that have been successful in one country can be applied elsewhere. The need for this is obvious if one considers the fact that today the variation between centers regarding sequence, technique, and timing of cleft repair could hardly be greater!

On the other hand, some obstacles, especially those arising from specific local circumstances and personalities, need specific local solutions. Among the difficulties mentioned were:

- Personal egotism of individuals unwilling to discontinue the practice of treating a few children each year;
- Competition between specialties for pre-eminence in the field, e.g., plastic-vs.-maxillofacial-vs.-pediatric-vs.-ENT surgery;
- Local pride, with every hospital, town, or region desiring its own small team or wishing to have local service;
- The necessity for teaching hospitals to cover a spectrum of clinical practice;
- Lack of responsiveness by the health authorities at local and national levels.

All of the above problems have confronted the UK in the recent past, and a national review was instigated by the government's Clinical Standards Advisory Group. The review included a national survey that revealed that Britain's fragmented, decentralized services were achieving a low standard of clinical care in some areas. As a result the Government instructed regions to provide care from a single regional center. Each of these should have a fully comprehensive specialist team, typically with two to three surgeons, each responsible for not less than 40 new personal cases for primary surgery per year. In this instance, government interest has been essential in trying to improve services when voluntary methods failed [16].

41.2.5 Subsequent Progress

Several changes in service organization across Europe subsequently occurred; e.g., some restructuring of services with more high-volume centers, improvements in the funding of treatment, attempts to improve record taking, and national registration with a view to intercenter research [22]. However, in some countries resistance to change is still evident. Reported obstacles include competition for patients between clinical disciplines, and a lack of collaboration between teams. Often, at the government level, cleft lip and palate care is not seen as a priority, and in some countries, difficulties with the insurance system and a trend towards fragmentation of services are evident.

41.3 EUROCRAN

EUROCRAN (European Collaboration in Craniofacial Anomalies), the final element of the Eurocleft initiative, began in 2000. It was conceived to harness and increase European research capability in cleft and related craniofacial anomalies. The project, funded by the

EU (Contracts No. QLG1-CT-2000-01019 and QLG1-CT-2002-30587) is designed to take forward several lines of research arising from the Eurocleft Cohort Study and related work in the Eurocleft Clinical Network. It involves a new partnership with a group of geneticists and epidemiologists assembled under a European Science Foundation program (No. GC/306 2/6/98, European Science Foundation, 2001). The project is designed to gain added value from the extended capability of the research group and from the increased potential for recruitment to clinical and genetic projects via the Eurocleft Clinical Network.

The project is supported by a website to facilitate dissemination and participation (www.eurocran.org), and consists of seven individual work packages, as follows.

41.3.1.1 A Group of Randomized Control Trials of Primary Cleft Surgery

A group of multicenter randomized trials of the primary surgery for infants with complete unilateral cleft lip and palate is testing four variations of surgical techniques in three concurrent trials. Infants have been randomized to a surgical method common to all three trials or the usual local method. A total of 450 infants is being recruited.

41.3.1.2 A Prospective Registry of Patients Undergoing Distraction Osteogenesis

This study has been carried out in two parts: the first is a web-based survey of the practice of distraction osteogenesis in Europe, the second a prospective registry of distraction cases with the aim of providing professionals with guidelines and recommendations for treatment and future research. One hundred and fifty patients have been registered to date, and the site can be accessed at www.eurocran.net.

41.3.1.3 Two Cohort Studies of Surgical Outcome in UCLP

A cohort of infants with unilateral cleft lip and palate (UCLP) at five centers is being registered with the study co-ordinating center prior to receiving the surgery normally practiced locally. At pre- and post-operative time points, specified data and clinical records are obtained. A second cohort of consecutively treated 5–7-year-olds with unilateral cleft lip and palate who

have previously received surgery are being recalled for standardized assessment. A comparison of peri-operative complications and operating costs will be made for cohort I and of dentofacial growth and nasolabial appearance for cohort II.

41.3.1.4 Development of a Good Practice Archive

An archive of clinical records for patients with unilateral cleft lip and palate treated consecutively in three centers in Europe representative of good practice has been assembled. The records include dental casts, photos, and cephalograms. This archive is a physical resource that is enabling cleft teams to monitor the quality of their care, and a number of cleft centers (Milan, Italy; Riga, Latvia; Tartu, Estonia) have already visited the archive to compare clinical outcomes as a series of pilot studies. Currently the archive is being further developed as a web-based library of clinical outcomes in order to facilitate comparison of clinical outcomes via the Internet.

41.3.1.5 A Gene-Environment Interaction Study

This multinational population-based case-parental triad study is seeking to investigate genetic susceptibility polymorphisms and gene-environment/gene-gene interactions operating in the etiology of orofacial clefting. Methodology includes a maternal interview on diet and other exposures in the periconceptual period, and DNA sampling of mother, father, and child leading to gene variant analysis. Over 1000 triads will be recruited to the study.

41.3.1.6 A Chromosomal Approach to Identifying Orofacial Clefting Genes

Patients with orofacial clefting associated with apparently balanced chromosomal rearrangements have been identified, and their breakpoints/clinical phenotypes have been catalogued. A bank of immortalized cell lines has been established from patients where two or more instances of a specific breakpoint has been associated with orofacial clefting (OFC). Genes that have been interrupted by two or more breakpoints have been identified. These genes are being fully characterized and screened for mutations and polymorphisms that may be used in the gene-environment interaction study [23].

41.3.1.7 Molecular Diagnosis of Monogenic Craniofacial Anomalies

Sensitive molecular assays for the mutations underlying a number of craniofacial malformation syndromes have been developed using Treacher Collins Syndrome (TCS) as a paradigm. This expertise is being disseminated to other molecular laboratories such that it will be available on a local basis [24].

41.3.2 The Global Context

In recent years there has been a growing recognition that international collaboration is a prerequisite for accessing adequate samples for research in the etiology, treatment, and prevention of craniofacial anomalies, and also for the assembly of a critical mass of clinical researchers and basic scientists in fields such as molecular biology, genetics, biochemistry, and epidemiology.

In 2000, the World Health Organization launched a five-year project designed to devise and disseminate an international research strategy. The work to date is reported in two publications [21, 25] (<http://www.eurocran.org/content.asp?contentID=1385&sid=145046>).

Those of us who have taken part in the European experiment described in this chapter recognize the challenges of international work. These include cultural and language barriers, though the latter has become a relatively minor problem for individuals fortunate to have English as a first language given its widespread use in the 21st century. The practicalities of rapid communication have of course been hugely enhanced by the advent of e-mail and teleconferencing.

Individuals' needs in demonstrating performance in research to their employing institution have been, for some, a deterrent to collaboration in the past. However this can be offset by participation in more substantial, adequately powered research projects than those generally arising in a local context, and by the emergence of new conventions of multiple authorship in leading journals.

Funding international research is a mixed challenge. The multicenter advantages of sample recruitment and skill mix can make for strong applications, but relatively few national funding agencies are keen to see too much of their budget flow beyond their national frontier. Strategies to promote international coordination of funding agencies are a high priority.

Craniofacial anomalies are a highly diverse group of complex congenital anomalies. Collectively they affect a significant proportion of the global society. As with many other aspects of the human condition,

there would seem to be many advantages to overcoming the challenges of international collaboration.

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42 Social, Ethical, and Health Policy Issues in the Care of Children with Major Craniofacial Conditions

RONALD P. STRAUSS

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42.1 Introduction

Craniofacial surgery and neonatal intensive care have made it possible for children with serious craniofacial conditions to live and often to experience effective habilitation. These therapeutic interventions also raise important social, policy, and ethical issues that are rarely examined. This chapter reviews the dilemmas which relate to the gatekeeper role for physicians, the impact of prenatal diagnosis, and the allocation of scarce fiscal and health resources to craniofacial care. The high degree of cost, the intense investment of medical resources, and the uncertain outcomes in the care of children with major craniofacial conditions must be considered in the distribution of resources within a health system. The rationing of health resources may be a future determinant of how care for major craniofacial conditions is delivered in the USA.

42.2 Craniofacial Treatment Decision-Making

"I felt shock, hopelessness and an overwhelming responsibility. With birth defects as bad as hers, it was hard to believe much could be done." – a 44-year-old mother of a 12-year-old child with Apert Syndrome.

The birth of a child with a major craniofacial condition is a serious crisis in the life of the family affected. Not only is the desire for a healthy baby unfulfilled, but the family must reframe their expectations for the child's needs. This may mean a re-evaluation of who is to provide care to the infant, how health needs will be met, and how much health insurance coverage is needed. The family may also need to consider the future health, social, and developmental limitations associated with their child's condition.

In the context of family crisis, clinical decisions must often be made about the extent, timing, and nature of the treatment which is to be provided. Family

values about the sanctity of life and norms for parental responsibility will guide decisions. Religious perspectives and ethnic or cultural values often provide the basis for making treatment decisions within the family [1–3]. Other social influences may affect decision-making and the available choices for parents and professionals.

Attitudes about disfigured children and about medical care may guide treatment decisions relative to children born with major craniofacial conditions. A fascination with disfigurement pervades mythology and Western literature [4, 5]. Combinations of fear, repulsion, interest, and sympathy affect attitudes about the child with a defect. Significant social ambivalence may be generated by children with obvious conditions. This ambivalence may find itself reflected in how a family or a clinician decides about major craniofacial surgery.

The child who is perceived as having a limited role as a future community participant or worker may be devalued, and treatment resources may be directed towards the needs of other children. In some settings, children seen as hopelessly afflicted may be neglected, maltreated, or abused [6]. In other sociocultural settings the child with a defect may be nurtured, seen as an exceptional child with special needs, and provided with the full range of possible treatment options. Historical and anthropological studies of child treatment and maltreatment [7] suggest that different cultures and different eras of time are characterized by markedly different attitudes and values placed on children and their health care needs.

Medical and societal resources and wealth will affect treatment decisions and options. In settings where treatment facilities are limited, as in many developing nations, the options for clinical activism may be severely curtailed. I am reminded of an experience with a rural Israeli mother who had just delivered a very premature, but otherwise healthy infant, in a regional hospital. The baby died soon after birth, there

being no neonatal intensive care unit (NICU) and limited medical expertise. The mother raised her head from the bed when I entered her room and asked me directly “would my baby have died if she were born in your country?” I knew the answer was that the baby would probably have been treated in the NICU and might be alive. How you might have answered her question will provide some insight into the complexities that surround treatment allocation. Medical facilities and health resources allow for care in previously untreatable conditions and create options for medical decision-making. They also raise new moral and ethical quandaries about who is to live and with what quality of life.

This chapter will examine the moral and health policy basis for making treatment decisions and will suggest social and ethical issues that may be raised in the care of children with major craniofacial conditions. It will consider four topics: (1) The History of Care for Children with Major Craniofacial Conditions, (2) The Gatekeeper Role for Physicians, (3) The Impact of Prenatal Diagnosis, and (4) Social Justice and Resource Allocation.

42.2.1 The History of Care for Children with Major Craniofacial Conditions

Until the last several decades many, if not most, children with major craniofacial conditions did not survive or experienced a much reduced quality of life [3, 8]. This occurred primarily because of the inability to provide nutrition, craniofacial surgical care, or respiratory assistance. Sometimes the lack of willingness to invest care giving or resources in the infant with a birth defect was influential. Infanticide [9, 10] and the withdrawal of caregiving or sustenance [11] have been discussed as possible responses to the infant with a marked defect. There is evidence that gender selection [12, 13] and the lack of skill in caring for such a child [14] may result in the infant's death. Infanticide is now universally seen as a crime, and most societies have developed social roles for children with birth conditions or defects. In spite of this, infanticide or child maltreatment may still occur hidden behind closed doors.

The success of neonatal intensive care units and craniofacial surgery has made it possible to treat many serious birth defects that may once have resulted in a child's death. It is likely that the application of these advanced forms of technological assistance have changed the profile of cases treated by many cleft/craniofacial teams. While craniofacial teams may once have predominantly seen children born with uncomplicated clefts of the lip and palate, many teams now report receiving referrals of children with

multiple defects and complicated craniofacial conditions [15].

This epidemiologic change is so substantial that many centers have changed their names from “cleft palate team” to “craniofacial team” [16]. With this marked shift in focus, clinicians may find themselves pondering difficult ethical questions. They may ask: should all children who have defects that might be repaired, receive treatment? Should infants with major and handicapping craniofacial conditions routinely be offered treatment in the NICU? Should they always get craniofacial surgical care? What criteria might be used to ration such care? Who should make the decision to treat or not to treat? To what extent does the physician serve as the agent of the society in directing the use of scarce and costly resources? In the context of a health care system, how many resources can be directed towards any individual child?

The advent of major craniofacial surgery, prenatal diagnosis, and neonatal intensive care raises basic social and ethical dilemmas relative to:

- a) Who should be treated and to what degree
- b) How scarce resources (i.e., medical or surgical services or expertise) should be allocated
- c) Whether and how cost/benefit calculations can be used in clinical decision-making
- d) What is the goal for major craniofacial surgery or treatment; and
- e) How much does the society value children and adults with disabilities.

The extremely high cost of correcting a complicated craniofacial condition [17] suggests that the society must consider the value of surgical care. Is the cost of repeated surgeries recouped in the development of human potential? Does the patient achieve social integration and productive vocational performance because he/she was treated? If not, what was the rationale of treatment? Do surgeons and families seek incremental improvements without regard to the final status and likely outcome of the process? Do surgeons have the capacity to manage access to their services? The role of the gatekeeper to medical and surgical services is a critical one, and the physician or the health insurer/managed care provider serves as an agent for the society in making treatment decisions.

42.2.2 The Gatekeeper Role for Physicians

The gatekeeper role [18, 19] involves the professional in exerting control over the availability of clinical treatment. Physicians encourage or discourage treatment based upon their legal responsibilities, values, and moral codes. Physicians are socialized during medical training to meet patients' needs and to deliv-

er a community service. They guide patients in accessing care and by making referrals when specialized expertise is required. The basic precept is to be of assistance, do no wrong, and to protect the public and community health.

Professional time and patient financial resources are not endless; thus health professionals serve to modulate and control access to their services, clinical skills, and time. Surgeons and other clinicians are constantly deciding whether it is worthwhile to undertake treatment, and thus they manage the market for their services. Physicians offer only treatment that they perceive as necessary and of benefit for the patient or the community. Often the surgical gatekeeper role is apparent when surgical treatment is denied. For example, a surgeon's decision not to operate on the palate of a non-verbal and developmentally delayed child until the child begins to speak is an application of the gatekeeper role.

The gatekeeper role may also relate to how a clinician rationalizes the investment of medical or surgical care. Social and psychological rationales for care may reflect the clinician's judgment that nonbiologic benefits may result from structural change. For example, the surgeon's decision to do a rhinoplasty on a fourteen-year-old girl with a cleft lip and minor nasal asymmetry may be based more on expected changes in her social experience rather than on her breathing ability. The use of psychosocial rationales for care is an application of the gatekeeper role.

The gatekeeping role allows physicians to manage many of the health care system's scarce resources, including the operating room and the Intensive Care Unit. Physicians determine who needs surgery and who requires highly technical hospital-based management. Insurance companies, managed care organizations, and other third-party payment agencies have developed mechanisms to monitor the use of these costly resources. Second surgical opinions, case managers, and insurance policy exclusions have routinely been used to control medical activism and expenditures. As the USA health care system continues to change and health care is delivered in a remarkably diverse and somewhat disorganized system, each patient and family may have a unique experience interacting with their payer, if covered at all. Physicians and other health professionals have expressed considerable dissatisfaction with the adversarial relationship that sometimes exists between the professional and the payer/insurer of care.

The responsibility of being a gatekeeper implies a considerable amount of control and power over one's work. Controlling the access to and use of one's own services may imply a conflict of interest. The gatekeeping role may be placed under stress when a physician is financially rewarded for being overly active

about care. When a surgeon who is charged with deciding how much care is necessary for a patient is also to receive financial reward from performance of the treatment, ethical roles may be stressed. This also may occur when outside parties such as insurance companies or managed care organizations are advocates for limiting the provision of care. They may gain from limiting the amount of service realized by a given patient or client.

In the matter of craniofacial clinical decision-making, the clinician serves as the agent of the society, making judgments which balance a variety of inputs. Factors which may play a role in these judgments include the likely benefits to the patient, the risks of treatment, the family's capacity to cope with the care of the child, the child's long-term prognosis, the cost of care, and the ability to access financial resources to pay for care.

The clinician's judgment may also relate to career goals and the mission of medicine. The physician may feel charged by society with the mission of "normalizing" children born with birth defects. Furthermore, the desire to perform dramatic and highly technical treatments on the "cutting edge" of medicine may be appealing. Both goals imply the likelihood of providing a patient with sophisticated and technical care. The desire to further the science and the clinical discipline by performing novel treatments may serve as a stress on the gatekeeper function.

Physicians primarily define their role in terms of the individual relationship with the patient and the family. Physicians in the USA are educated to see themselves as advocates for the child patient as well as the parents, and have difficulties in moments where the interests of patients and parents may not coincide. For example, in a situation where a child has a serious defect that would require intense and costly care, and the family expresses a wish to limit treatment, the clinician may be placed in a difficult position. Whose interests does the physician protect? Who advocates for the child's interests? Does the tendency to medically intervene generally prevail? Do the parents always have the final say relative to a treatment decision? In recent years legislative and judicial decisions have become central in guiding medical decision-makers and in defining the limits of medical autonomy (e.g., the "Baby Doe" cases) [20].

Medical education socializes Western physicians to be activist in their clinical responses. In simple words, due to their training, doctors try to fix disabilities when possible. The decision not to offer care, or to withdraw care already started, is always difficult and places the physician in struggle with well socialized values to heal. In new or innovative forms of treatment, few guidelines or norms may exist for deciding about whether to intervene. In cases of craniofacial

surgery, activist principles and the desire to fix disabilities predispose the physician to providing treatment whenever possible.

Periodically in the life of a profession, specific technological innovations markedly alter the course of medical practice and thinking. This occurred with the introduction of the stethoscope, aseptic technique, and the use of antipsychotic medications. This also occurred with the introduction of prenatal diagnosis and the introduction of craniofacial surgery. The introduction of these technologies has markedly changed how medicine deals with congenital disabilities. Prenatal diagnosis and ultrasound imaging have created a situation where parents have the option of whether to bear a child with a known defect. Craniofacial surgery has created the option to correct defects that may exist. Parents must weigh the pain of bearing a child with a birth defect and the expectation of surgical correction against their moral values and may face the choice of whether to abort the child. Craniofacial surgery and prenatal diagnosis are both possibly major components in the family and parental decisions about the child with a major craniofacial condition.

42.2.3 The Impact of Prenatal Diagnosis

Prenatal diagnosis and imaging of major craniofacial conditions is increasingly common [21–23]. The use of ultrasound allows for the early visualization of fetal defects. Techniques such as amniocentesis, genetic screening, and risk appraisal may provide the family and physicians with information about a child's craniofacial condition prior to birth. The knowledge of a condition during the prenatal period implies the possibility of choice relative to the birth of a child with a defect.

Knowledge about defects prior to birth has caused medicine to face profound questions about how we as a society will deal with prenatal information about serious craniofacial conditions. Will there be significant pressure on mothers to abort such fetuses in order to avoid the cost and pain of corrections? Will there be rewards for parents who decide to save the society the cost of treatment? Will we stigmatize and harass parents for seeking to terminate a life, regardless of quality, by abortion?

The realities of the high cost associated with the survival and treatment of children with major craniofacial defects may affect how we perceive a parent's decision to abort a fetus with an identified condition. A decision to bear such a child implies major expenditures for the parents and/or the society.

Advocates against the availability of abortion in the USA will eventually clash with the reality that some

parents of fetuses with significant conditions may wish to terminate the pregnancy. Indeed, prenatal diagnostic technology is based on the premise that some action may occur in response to an untoward finding on testing. Some parents may merely be seeking prenatal information to be prepared for the birth of their child. Other parents may want to arrange for adoption of a special-needs child, while still others will seek abortion. Social policy debates will determine whether abortion remains available or becomes illegal. The debate may consider whether society is willing to pay the costs of maintenance and rehabilitation of congenitally disabled children and adults.

Social values relative to aesthetic and functional conformity are also challenged by prenatal diagnosis of birth defects. Will infants who are different or have birth defects be aborted or be accepted? How major a condition must there be to rationalize an abortion? Will there be less tolerance of different appearance or identity if those with different identities are aborted or corrected? To what extent does living with a range of appearances and disabilities humanize a society? Some would argue that medicine should exert itself to make children look as normal as possible to meet social expectations. Others would say that instead of changing the child or his appearance, the focus should be on changing social values to encourage the acceptance of those who appear different. For the moment, there is little question that those who look different are treated in Western societies as less than equal and often are stigmatized.

The introduction of the current array of sophisticated prenatal screening mechanisms did not imply that medicine was prepared for the social and ethical ramifications of their use. Similarly, phenomena such as fetal surgery and neonatal intensive care pose ethical dilemmas. One would hope that the introduction of new biomedical technologies implies the willingness to consider the complex issues these procedures raise for clinicians and for those engaged in health care financing and policy.

42.2.4 Social Justice and Resource Allocation

It is sometimes argued that when limited resources exist, health dollars are best spent on prevention, rather than treatment. Some would state that health and economic resources are most rationally invested in maternal health care and in the application of established preventive regimes. Thus, it could be argued that prevention of fetal alcohol syndrome is preferable to focusing resources on its treatment. Most would agree that when possible, a preventive approach is preferable.

The USA health system, however, often functions to support treatment, as opposed to preventive, approaches. In the case of cancer and heart disease, known risk factors might be reduced, yet the bulk of health dollars are spent on the treatment of existing conditions. In the case of major craniofacial conditions, resources might best be spent on research and on the prevention of defects. This might involve developing programs that encourage maternal health and nutrition, genetic counseling and screening, as well as prenatal ultrasound/diagnosis. A reasonable approach to health policy might be built on research into craniofacial health promotion and disease prevention.

However, the major craniofacial disorders are rare and currently are often of unknown etiology. Given the paucity of preventive craniofacial efforts, the bulk of resources are currently focused on the clinical treatment of these conditions. In the face of the reality of a child who is born with a major craniofacial condition, all attention is understandably drawn to the clinical treatment possibilities.

Physicians are not socialized to ask questions about social justice when faced with a specific patient's needs. Few would find it reasonable for a clinician in this situation to worry about whether the society's resources are better spent on public health or immunization rather than on expensive surgical reconstruction. Most citizens would agree that public dollars would be better invested in the realm of prevention, and yet when faced with the denial of reconstruction to the individual patient due to limited resources, the social will to persevere diminishes.

Under circumstances of individual patient need, it is difficult to pose questions about the appropriate allocation of economic and clinical resources or social justice. In the context of ethical discussion, one may consider questions of fairness and equity.

Values regarding the sanctity of the individual are strongly held in the USA, and rationing of health care has proven to be a difficult prospect [24, 25], although such rationing occurs in a *de facto* way regularly. When rationing of health care does occur, it often is due to limited insurance or managed care policies, total lack of health coverage, or because of a scarcity in providers or hospital facilities. For the individual clinician these barriers to care may not be apparent, since many patients who reach his/her door are able to pay for care. The issues arise when payment is not available and when care is very costly. If patients cannot themselves afford care, and insurers are unable or unwilling to pay, then the true impact of rationed clinical care becomes apparent.

Health care is already allocated in the USA. Low SES and minority populations do not uniformly have equal access to high-quality health services in the

USA. This is reflected in their relatively lower health status compared to more affluent or non-minority populations [26]. In a society where marked disparities in access to health care exist, one must ask whether the ability to pay is the most just medium for deciding who gets care. In the case of major craniofacial conditions, should ability to pay for expensive services guide their availability?

In the USA, insurance carriers, managed care organizations, case managers, or governmental agencies may determine who receives medical care. What values guide their decisions? Do they seek to provide maximum good to the largest number of citizens? Do they seek to provide a basic level of good to all? Do they seek to meet the demands of special groups of needy patients? In determining benefits, who speaks for the interests of children? In the case of costly procedures that benefit very few persons, political advocacy seems unlikely to be a large force in guiding insurance coverage. Principles of social justice do come into play when decision-makers consider how much pain would occur if care were denied, or how much gain can be achieved by funding treatment. These distinctions are difficult, and cost/benefit thinking may be used in making such funding decisions.

Decisions about making extraordinary expenditures on highly technical craniofacial surgery and care may be deliberated by health agencies and insurers. In group policies, particularly those which are self-insurance or small business programs, the cost of catastrophic illness or a major birth defect in a single member or child may affect the entire group and its cost of insurance. There are many examples of health insurance programs that seek to avoid expensive claims or that withdraw continued coverage from the neediest claimants.

In calculating coverage and policies, insurance companies are not likely to consider how the cost of a craniofacial repair might compare with the costs of maintaining a person's disability. Such companies seek to minimize their costs, and are not responsible for dealing with the long-term social consequences of a disability. Governmental agencies, on the other hand, may ask questions about long-term patient function. They may consider whether the person will be self-supporting and self-maintaining, in the long term, after care. A decision to provide costly care to a child may reflect the perception that treatment will reduce the burden to society of lifelong assistance or maintenance.

Another approach to cost-benefit considerations recognizes the social cost of living around deformed persons, who may inadvertently challenge social norms of acceptability. Some conditions elicit negative social responses that provoke society to cover up the difference or reduce the deviance. It is in this fash-

ion that significant social pressure may actually be placed on the person with a major craniofacial condition to participate in and agree to reconstruction. The person is expected to "be a good patient" and seek amelioration.

Perhaps the most difficult resource distinction regards the relative worth of social investment in various diagnoses. Why is a cleft palate repair seen uniformly as cost-effective, while some question a Kleeblattschädel craniosynostosis repair? Is it that we expect a better outcome from the cleft surgery? Or is it that the frequency of clefts demands their repair, while the scarcity of major craniofacial syndromes does not? If we can afford as a society to provide cleft repairs, why then not afford craniofacial repairs? Clearly questions of the magnitude of cost exist. The costs as calculated cannot be merely the direct costs of several years of surgical care; rather they must also include the costs of parental work loss, of special education, of rehabilitation services, of mental health assistance, of lost patient productivity, and of mental anguish. These costs are generally immeasurable, yet they constitute real components of the losses realized.

In the current context of limiting health dollars and resources, it is predictable that some rationing decisions will occur. The deciding principles to be applied are unclear, but they will determine who will live, and who will live well; who will be cared for and who will languish; who will receive benefit and who will suffer. These distinctions will directly affect how care for major craniofacial conditions will be delivered in the United States and other developed nations.

42.3 Justice and the Distribution of Resources

This chapter has sought to review the social and ethical ramifications of craniofacial surgery for children born with major craniofacial conditions. The remarkable tools of craniofacial surgery and neonatal life support/intensive care have made it possible for many more children with serious defects to live. Their survival has raised questions about how much a society is willing to invest in their care, especially considering the impact that even a repaired defect will have on the patient's quality of life. The availability of prenatal diagnosis and abortion may present additional choices to parents and clinicians. After a birth of a child with a major birth defect, the intense expense and the high degree of clinical activism involved heighten our perception of the costs of craniofacial surgery. The desire to utilize new techniques, to advance a discipline, and provide patient benefits may affect clinician decisions. In the context of these often conflicting social currents, many questions about justice and the

distribution of resources arise [27]. Should society invest large amounts of effort and resources in costly corrections of rare and disabling conditions, or should it spend limited health dollars on population-based preventive efforts? [28, 29] Decisions about rationing care will arise in the context of limited resources and in societies where demands on the health system exceed the system capacity. Craniofacial care poses many social, ethical, and health policy issues [30–36] for health professionals, parents, and the community-at-large.

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Subject Index

A

- Abbe flap 470, 518
- Aberrant muscle force 43
- Abnormal growth 10
- Accutane 290
- Achondroplasia 290
- Acoustic bridge 355
- ACPA see American Cleft Palate-Cranio-facial Association
- Acrylic palatal vault 453
- Acrylic resin 398, 726, 727, 728, 731, 732
- Adaptol 731
- Adenoid 647, 649
 - morphology 668
 - size 634
- Adenoidectomy 52, 660, 669, 677, 709
- Adenoid mass 52
- Aerodynamic testing 710
- Aesthetic facial balance 525
- Aesthetic scale 610
- Air flow assessing device 741
- Airway deficiency 657
 - maintenance 286
 - obstruction 221, 519
- Alar cartilage 463
 - framework 472
 - suspension 461
- Alar marginal incision 461
- Alar rim 452, 456
- Alginate 243, 244
- Allergy 265
- Allograft 596
- Alpha-smooth muscle actin see ASMA
- Alternate Rapid Maxillary Expansions and Constrictions see ALT-RAMEC
- ALT-RAMEC (Alternate Rapid Maxillary Expansions and Constrictions) 537, 539, 541, 542, 549
- Alveolar bone 603
- Alveolar bone graft/grafting 59, 72, 84, 91, 96, 118, 207, 460–462, 525, 540, 544, 547, 548, 575
 - secondary 111, 118, 119, 149, 151, 158, 162, 163, 166, 179, 208, 323, 351, 364–366, 409, 414, 432, 444, 482, 484, 488–490, 507, 512, 758
- Alveolar bone growth 137
- Alveolar bone support 133
- Alveolar cleft 548
 - deformity 452
 - space 80, 382
 - secondary bone grafting 601
- Alveolar cranial bone graft 76, 82, 91
 - secondary 123, 129, 132, 143
- Alveolar crest 481, 603, 605
- Alveolar deficiency 274
- Alveolar fistula 524
- Alveolar labial plate 332
- Alveolar osseous base 593
- Alveolar periosteoplasty 279
- Alveolar process 63
- Alveolar ridge 364, 377, 383, 401, 447, 453, 474, 592, 593
 - analysis 343
 - optimum position 383
- Alveoloperiosteoplasty 398
- Alveolus 46, 55, 56, 57, 58, 59
- Ambryonic gut tube 4
- American Association of Oral Surgeons 276
- American Association of Plastic Surgeons, Chicago 276
- American Cleft Palate Center 343
- American Cleft Palate Craniofacial Association (ACPA) 285, 416, 459, 488, 691, 694, 705, 777
 - meeting St. Louis 391, 395
 - meeting Tampa 385
- American Institute of Ultrasound in Medicine 14
- Amyotrophic lateral sclerosis 740
- ANB angle 83
- Aneuploidy 14, 15
- Angle of convexity 274
- Angular changes 522
- Angular gyrus 711
- Ankyloblepharon 203
- Ankyloglossia 203
- Anoxia 274
- ANS (anterior nasal spine) 522
- Antepartum obstetrical ultrasonographic evaluation 14
- Anterior alveolar ridge 363
- Anterior arch discrepancy 127
- Anterior bony apposition 759
- Anterior cleft
 - closure 323
 - space 129, 130, 136, 137, 153, 154, 156, 163, 175, 185
- Anterior cranial base 18
 - growth 123, 186
- Anterior crossbite 124, 128, 488, 489, 509, 512, 522, 528, 602
 - correction 94
- Anterior displacement of maxilla 536
- Anterior nasal spine see ANS
- Anterior palatal oronasal opening 151
- Anterior removable prosthesis 78
- Anterior-posterior arch length 433
- Anteroposterior crossbite 591
- Anteroposterior maxillary development 382
- Anteroposterior maxillomandibular discrepancy 525
- Anxiety 258
- Apert syndrome 268, 287, 530, 635, 777
- Apoptosis 303
- Armamentarium of pharyngeal flap 503
- Arnold expander 142, 204, 205
- Articulation
 - age 696
 - competence 607
 - development 698, 708
- Articulatory therapy 712
- Ascending ramus 555–557, 561, 563, 569
- ASMA (Alpha-smooth muscle actin) 303
- Atlanto-axial dislocation 628
- Atlanto-dental angulation 630
- Attractiveness, studies on 258

Audiology 289
 – early identification 289
 – management 289
 – monitoring 289
 Auditory brainstem response testing
 355, 356
 Auditory canal, external 357
 Auditory system 355
 Auditory training 639, 640
 Autogenous bone 180
 – graft/grafting 507, 512, 525, 530
 Autogenous cancellous bone 587
 Autologous cancellous bone 604, 605
 Autologous cartilage graft 676
 Autopolymer 731

B

Babbling 711
 Basic occipital bone 623
 Basion horizontal 18, 19, 29, 31
 – concept 28
 – facial polygon 131
 – method (Coben) 76, 83, 90, 93, 141, 158, 172, 186, 341, 484
 – procedure 439, 445
 Bayley Scales of Infant Development see BSID
 BCCLP (bilateral complete cleft lip and palate) 226, 227, 229, 230
 BCL (bilateral cleft of the lip) 317
 BCLP (bilateral cleft of the lip and palate) 26, 27, 31, 49, 50, 107, 108, 142, 159, 163, 172, 173, 201, 202, 208, 210, 238, 239, 322, 327, 330, 351, 387, 389, 397, 398, 431, 452, 487, 495–497, 521, 528, 540, 547, 575–577, 579, 580, 660, 664, 696, 708
 – unoperated 247, 248
 – variations 50
 Behavior problem 257
 Bergen Cleft Palate-Craniofacial Center 488, 491
 Bergen rationale 487, 491, 493
 Berkowitz and Millard palatal cleft closure timing 344
 Berkowitz study 755
 Berkowitz's cephalometric and nasopharyngoscopic studies 52
 Berkowitz's clinical report 329
 Berkowitz's observations 753
 Berkowitz's serial growth study 342, 385
 Berkowitz's serial occlusal study of CICLP 342
 Bifid uvula 53
 Bilateral alveolar cleft 48
 Bilateral cleft lip nose 273
 Bilateral cleft of the lip and palate see BCLP
 Bilateral complete cleft of the hard and soft palate 646
 Bilateral posterior crossbite 494
 Bilingual family 712
 Bimaxillary protrusion 252
 Bimaxillary retrognathia 227, 229, 233
 Bimaxillary retrusion 249
 Biocompatible synthetic membrane 307
 Biofeedback therapy 618
 Biomedical technology 780
 Biostereometric technique 22
 Bipedicle island flap 515, 516
 Bird face deformity 561, 562, 569
 BMP cytokine 587, 596
 Bolton standards 106
 Bone Autotransplantation 604
 Bone graft/-ing (s. also Autogenous bone grafting) 66, 599
 – material 596
 – primary 279
 – secondary 279
 – surgical procedure 592
 Bone manipulation 599
 Bone remodeling 32
 Bone vascularity 568
 Boneless bone grafting 278
 Bonferroni procedure 365
 Bony ankylosis 341
 Bony alveolar bridge 423
 Bony overgrowth 102
 Bony pharynx 65
 Bony union across alveolar cleft 551
 Breast malignancy 280
 Bridgework 605
 Broad personality type 257, 258
 Brogan technique 398
 Brophy method 396, 448
 BSID (Bayley Scales of Infant Development) 715
 Buccal alveolar surface 318
 Buccal and anterior occlusion 331
 Buccal crossbite 84, 109, 112, 120, 121, 124, 146, 162, 330, 337, 447, 503, 509, 512, 602
 – correction 203, 204, 205, 206, 207, 208
 Buccal flap 593
 Buccal mucosa 302
 Buccal occlusion 56, 88, 169, 333, 348, 481
 – class I 161, 413
 – class III 414, 418
 Buccal sulcus (Burian) 512
 Buccinator 43, 44
 Bulbar polio 637, 740

C

Calvarium 463
 Cancellous bone 592
 Cancellous iliac crest bone 105
 Canine tooth 593

Caries 209, 325
 Carotid artery 669
 Carpenter syndrome 287
 Carry effect 315
 Cartilage implant 641
 Cartilage intercellular matrix 451
 Cartilaginous growth 28
 Cartilaginous septum 104
 Catch-up growth 329, 342, 344
 Caucasian population 706
 CBCLP (complete bilateral clefts of the lips and palate) 28, 37, 39, 44, 45, 47, 99, 100, 102, 103, 105, 111, 113, 114, 124, 132, 151, 173, 210, 316, 321, 323, 328, 343, 344, 364, 365, 367, 374, 376, 383, 386, 389, 398, 409, 410, 414, 415, 418, 428, 440, 441, 443–446, 449, 470, 479, 484, 751, 755, 756, 760–762
 CDI (Communicative Development Inventory): Toddler 714
 Cell enhancement through tissue culturing 599
 Cellular destruction 103
 Cellular proliferation 325
 Cephalometric evaluation 521
 Cephalometric laminagraphy 51
 Cephalometric measurement 17
 Cephalometric roentgenogram 49, 215, 222
 Cerebral vascular accident 650, 740
 Cervical periodontal ligament 305
 Cervical spine anomaly 628–631
 Cervical vertebrae 21, 622
 Chewing 711
 Chicago Dental Institute 276
 Child
 – adjustment 259
 – early communication attempts 697
 – overcontrolled 258
 – undercontrolled 258
 Children's Hospital of Philadelphia 280
 Children's Memorial Hospital Cleft Palate Clinic, Chicago 297, 397
 Chin disproportion 582
 Choanal atresia 286
 Cholesteatoma 360, 361
 Chondrocostal grafting 569
 Chondrocranium 26
 Chromosomal abnormality 14, 15
 Chromosome 22-deletion DiGeorge syndrome 5
 Chromosome 22q11.2 deletion 673
 Cinefluoroscopy 616
 Cineradiographic study 721
 Circumzygomatic wire 514
 CL (cleft lip) 48, 55–59
 – inheritance 52
 – isolated 227, 230
 – repair 588
 – rhinoplasty 470, 518
 – surgery, secondary 188

- CL with or without cleft palate 290
 - chromosomal testing 290
 - multifactorial inheritance 290
 - prenatal diagnosis 290
- Cleft buccal segment 69, 74
- Cleft Center 684
- Cleft embryo-pathogenesis 487
- Cleft lip see CL
- Cleft lip and palate see CLP
- Cleft maxillary hypoplasia 521
- Cleft of the uvulae 55, 59, 60
- Cleft orbicularis oris muscle 102, 315
- Cleft palatal segment 44
- Cleft palate see CP
- Cleft Palate Center of Amsterdam
 - Free University Hospital 400
- Cleft Palate Center of Nijmegen
 - Universty Medical Center 400
- Cleft Palate Center of Rotterdam
 - Erasmus University Hospital 400
- Cleft Palate Centre of the University of Illinois 215, 218, 219
- Cleft Palate Institute of the North-western University Dental School 384
- Cleft Palate Team at New York University
 - Medical Cente 399
- Cleft palate only see CPO
- CP (cleft palate) 48
 - closure 320, 363, 461
 - growth 316
 - inheritance 52
 - isolated 50
 - orofacial growth 383
 - prosthesis 723
 - speech 691, 700
 - submucous 52, 53
 - surgery 307, 308, 719
 - surgical history 344
 - team 682, 683
 - timing 363ff
- CP alone
 - chromosomal testing 291
 - molecular (DNA) testing 291
- CP-craniofacial team 607
- Cleft research 751
- Cleft subject 364
- Cleft surgeon 573
- Cleft surgery 753
- Cleft team 259
- Cleft, submucous 285
- Cleftal bone
 - defect 596
 - grafting 587
- Clefting process 210
- Cleft-orthognathic surgery 573
- Clinical practice 772
- Clinical research 692, 751
 - scientific evidence 692
- Clinical Standards Advisory Group 773
- Clinical trial 20
- Close the hole 319
- CLP (cleft lip and palate) 226, 229, 230, 233, 237, 257–259, 487–489, 495, 698, 713, 715
 - relationship to brain structures and function 259
 - treatment 491
- Coben's basion horizontal analysis (s. also Basion horizontal) 65
- Coben's growth philosophy 19
- Cocaine abuse 462
- Cognitive development 258
- Cohort study 774
- Collagen 676
 - type I 304, 305, 309
 - type III 303, 304
 - type IV 4
- Collito-Walker technique 317
- Columella 49, 451, 452, 455
 - absent 451
 - deficient 451
 - elongation 474
 - nonsurgical elongation 453
 - nonsurgical lengthening 455, 461
 - oblique 451, 452
- Combination prothesis 738, 739, 740
- Communication
 - development 695, 699, 700
 - disorder 678
 - failure 692
 - impairment 615, 716
- Comparative growth study 375
- Complete bilateral clefts of the lip and palate see CBCLP
- Complete nasal obstruction 634
- Complete rotation advancement 468
- Complete unilateral cleft lip and palate see CUCLP
- Concave facial profile 418, 427, 524
- Congenital palatal insufficiency see CPI
- Consensus recommendations 771
- Contiguous anatomy 349
- Contiguous skeletal structure 210
- Continuous phonation 735
- Continuous positive airway pressure see CPAP
- Conventional osteotomy surgery 570
- Convex facial profile 444
- Core curriculum 283ff
- Cortical bone 601
- Corticotomy 556, 557, 560, 561, 569
- Cortisone 7
- Cost-effectiveness 400, 403, 460
 - ratio 487
- CP (cleft palate) 230, 233
- CPAP (continuous positive airway pressure) 673
 - therapy 711, 712, 717
- CPI (congenital palatal insufficiency) 53, 628, 629
- CPO (cleft palate only) 713, 715
- Cranial base abnormality 635
- Cranial bone graft 70, 505
- Cranial cartilaginous tissue 27
- Cranial cell migration, disruption 259
- Cranial floor 24
- Cranial neural crest 4
- Cranial stabilization 530
- Craniofacial anomalies 285, 286
- Craniofacial biology
 - basic definitions 288
 - developmental 287
- Craniofacial building block 3
- Craniofacial care 782
- Craniofacial Center at the Univesity of Illinois College of Medicine 762
- Craniofacial Center of the Hospital for Sick Children 64
- Craniofacial cleft 287
- Craniofacial complex 325
- Craniofacial disjunction 566
- Craniofacial embryology 287
- Craniofacial growth 18, 28, 63, 233, 304, 558, 571, 697, 749
 - direction 17
 - postnatal 28, 29
- Craniofacial morphogenesis 3, 225
- Craniofacial morphology 227
- Craniofacial psychology 263ff
- Craniofacial scoliosis 555
- Craniofacial skeleton 555, 559
 - remodeling 555ff
- Craniofacial surgery 749, 777, 778, 780–782
- Craniofacial synostosis 32
- Craniofacial treatment, decision-making 777
- Cranio-maxillary complex 28
- Craniomaxillofacial fixation 577
- Craniomaxillofacial skeleton 530
- Craniosynostosis 361
- Craniosynostosis 286, 287, 555, 564, 568, 570
 - syndromal 287
- Cribiform plate 625
- Crouzon syndrome 287, 290, 361, 530, 567, 635
- CUCLP (complete unilateral cleft lip and palate) 22, 28, 31, 37, 38, 44, 47, 61, 62, 64, 66–68, 79, 84, 89, 94, 95, 210–212, 241, 243, 318, 319, 337, 341, 343–345, 350, 364–367, 374, 376, 383–387, 391, 398, 409, 410, 414, 415, 417, 418, 425, 427, 434, 436, 437, 444, 448, 449, 479, 481, 490, 524, 751, 754–756, 761, 762, 765
- Cul-de-sac shifting 738
- Cultural background 267
- Cupid's bow 273, 274, 275, 276, 318, 461, 470
- Cushing elevator 510
- Cuspid crossbite 63, 166, 183, 346, 347, 348, 481
- Cuspid substitution 462
- Cyanosis 215
- Cytokines, pro-inflammatory, pro-fibrotic 302

D

Dahl's Danish group 341
 Danish study of craniofacial morphology 229
 Dautrey osteotomy 510
 Deciduous dentition 118
 Decreased shelf forces 753
 Deep pharynx 647
 Deficiency in lateral wall movements 52
 Deficient soft palate 723
 Degenerative central nervous system disease 740
 Deglutition 396
 Delaire-style protraction facial mask 91, 93, 111, 480, 428, 602
 Delayed surgery 724
 Dental adjustment 491
 Dental changes 523^c 525
 Dental crossbite 344
 Dental crowding 338, 427
 Dental defect 46
 Dental dysplasia 207
 Dental malocclusion (s. also malocclusion) 209, 489
 Dental occlusal and facial study 409
 Dental occlusion 113, 326, 348, 363, 409, 417, 441, 487, 555, 557
 – ideal 86
 Dental overbite 127
 Dental overjet 127
 Dental protrusion 32
 Dental reconstruction 177
 Dental stone 453, 728
 Dental study model 238, 239, 240, 241, 249
 Dentition 201, 204, 301,
 – permanent 206, 207
 Dentition 301
 – aberrant 524
 – deciduous 206, 207, 209, 326, 351, 375, 418, 495, 496
 – mandibular 538
 – maxillary 496
 – mixed 118, 207, 209, 479
 – mutilated 580
 – permanent 207, 375, 418, 489, 495, 602, 604, 727
 Dentoalveolar growth 329
 Dentoalveolar mandibular proclination 488
 Dentoalveolar process 304
 Dentoalveolar proclination 495
 Dentoalveolar prominence 540
 Dentoalveolar relapse 602
 Dentofacial complex 17
 Dentomaxillary growth 304
 Dento-osseous-musculo-mucosal flap 577
 Denture prosthesis 720
 Detrimental psychosocial ramification 524

Developing/least developed world 237
 Developmental critical period 8
 Developmental field concept 5
 Developmental malformation 268
 Developmental reading disability 259
 Developmental study 752
 Deviated septum 662
 Diagnosis-related checklist 497
 Differential diagnosis 342, 344, 363, 376
 DiGeorge syndrome 5, 715
 Disability group 263
 Discrimination 263
 Distorted neonatal cleft arch form 35
 Distorted nostril 47
 Distraction osteogenesis 111, 418, 463, 529, 530, 535, 540, 555ff, 569, 570, 573, 582, 774
 Donor cavity 601
 Donor site 604
 Donor-site morbidity 571
 Dorsal position of the tongue 396
 Dorsiflexion 221
 Double-hinged expander 539
 Double-hinged rapid maxillary expander 536, 537
 Down syndrome 280, 716
 Downward displaced premaxilla in bilateral cleft patients 540
 Duoderm 457
 DUTCHLEFT Study 400–404, 699
 Dynamic sphincter 672, 673
 Dyslexia, 259
 – dysnomia model 258
 Dysmorphogenesis 10

E

Ear
 – pneumatic system 355
 – reconstruction 286
 Early facial morphogenesis 7
 Economic health evaluation 403
 Ectodermal dysplasia 530
 Ectopic tooth eruption 88, 207
 Edentulous maxilla 530
 Effective maxillary orthopedic protraction 535
 EGF (epidermal growth factor) 302
 EGFR 9
 Elastic traction 35
 Electromechanical digitizer 129, 136
 Electromyographic (EMG) activity 711
 Emotional self-regulation 257
 Emotional adjustment 259, 266
 Encephalitis 650
 Endoscopic optical system 355
 Endotracheal anesthesia 566
 Epidermal growth factor see EGF
 Epiglottis 216, 217, 617
 Epilepsy 265
 Epinephrine/hyaluronidase solution 503, 510

Epithelialization 329, 750, 752
 Equine appearance 540, 541
 Esophagus 621
 Estrogen
 – level 452
 – maternal 460
 Ethical Committee 400
 Ethical guideline 21
 Ethmoid bone 211, 625
 Eurocleft Clinical Network 765, 771, 773, 774
 Eurocleft Cohort Study 765
 Eurocleft Policy Statements 771, 773
 Eurocleft Practice Guidelines 771, 773
 Eurocleft studies 280
 EUROCRAN (European Collaboration in Cranofacial Anomalies) Research Programme 765, 770, 773, 775
 European cleft palate center 343
 European Collaboration in Cranofacial Anomalies see EUROCRAN
 European research capability 773
 European services 771
 Eustachian tube 243, 244, 355, 357, 358, 359, 361, 647, 674, 732
 – patency 356
 – test 360
 Excessive scar tissue 457
 Exogenous cytokine 599
 Exophthalmos 361
 Expansion prosthesis 724
 Expiratory phase 662
 Extensive mucoperiosteal undermining 207
 External cervical root resorption 604
 Extraction of mandibular teeth 489
 Extraoral anchorage 451
 Extraoral strapping 395

F

Face
 – flattening 64
 – mask distraction groups 523
 – mask elastic distraction 522, 523, 529
 Face-to-face interaction 265
 Face-to-face professional interaction and discussion 770
 Facial aesthetics 512, 326, 343, 363, 487, 497
 Facial and palatal growth study 326
 Facial angle 18
 Facial appearance 490
 Facial bone growth 274
 Facial cleft 13
 – antenatal diagnosis 13
 – deformity
 – – severity 266, 267, 268
 – – self-perception 267
 Facial concavity 527
 Facial deformity 263

Facial development of cleft children 225
 – iatrogenic factors 225
 – adaptive factors 225
 Facial force diagram 35
 Facial grimacing 682
 Facial growth 21, 23, 79, 84
 – direction 28
 – malformation 325
 – pattern 76, 449, 749, 752
 – rotations 31
 – standards 64
 – studies 113, 105, 325
 – – longitudinal 316
 Facial landmark 18, 19
 Facial morphology 226
 Facial occlusion 441
 Facial polygon 18
 Facial profile evaluation 433
 Facial protraction mask 420
 Facial scar 265
 Facial skeletal surgery 749
 Facial surgery 530
 Facial symmetry 557
 Facial-palatal growth study 185
 – longitudinal 329
 Facioscapulohumeral muscular dystrophy 650
 Failure to fuse 753
 Family anamnesis 488
 Family crisis 777
 Faradization 740
 Fascination with disfigurement in mythology and Western literature 777
 Faulty learning 626
 Feeding 286
 Fetal alcohol syndrome 780
 FGF (fibroblast growth factor) 4, 302, 303, 309
 FGFR (Fibroblast growth factor receptor) 288
 Fibrinogen 302
 Fibroblast growth factor see FGF
 Fibroblast growth factor receptor see FGFR
 Fibroblast
 – dermal 309, 310
 – fetal 309
 – mucosal 310
 – oral 309
 Fibronectin 4, 302, 303
 Fibrosis 103
 Fibrous bands of scar tissue 325
 Final occlusion 565
 Finnish-speaking child 708
 First International Congress on Plastic Surgery, Stockholm 275
 Fishmouth 670
 Fistula 202, 204, 547
 – closure 525, 577
 – formation 349
 Fixed appliance therapy 602

Flexible fiberoptic tube 617
 Florida Cleft Palate Association 459
 Foramen magnum 19, 29
 Forced-response test 360
 Fork flap procedure 165
 Forward-growing mandible 444
 Frankfor horizontal plane 18, 19, 615, 617
 French nose 568
 Frenum attachment 456
 Friede and Pruzansky's cephalometric report 107, 108
 Fronto-nasal prominence 7
 Fronto-orbital advancement 564
 Fronto-orbital area 566
 FSIQ (full scale intelligence quotient) 258, 259, 267
 Full dental rehabilitation 605
 Full scale intelligence quotient see FSIQ
 Function Matrix Principle 23, 24
 Functional distortion 301
 Furlow double opposing Z-palatoplasty 667, 462, 640, 673-675

G

Gatekeeper role 778, 779
 Gene therapy 599
 Gene-environment interaction study 774
 General dentist 573
 Genetic Control Theory 23
 Genetic engineering treatment method 599
 Genetics 290
 Genioplasty 575, 578
 Genitourinary system 203
 – anomalies 203
 Geometric displacement 44
 Gingival mucoperiosteum 302
 Gingival recession 550
 Gingivitis 209
 Gingivoperiosteoplasty (GPP) 38, 112, 279, 400, 411, 417, 460, 461, 463, 468–470, 547, 551
 Global context 775
 Glossoptosis 215, 218, 219, 221, 222, 227
 Glottal stop substitution 740
 Goldenhar's syndrome 555
 Good Practice Archive 769, 774
 GOSLON yardstick 238, 245
 Goteborg Team 384
 GPP see gingivoperiosteoplasty
 Great Debate 395
 Greatest esthetic challenge 451
 Growth
 – acceleration curve 350
 – center 396
 – deficiency 381, 391
 – disturbance 301, 398, 400
 – hormone secretion 252

 – retardation 349, 750
 – studies 218, 220, longitudinal 221
 – velocity 368, 369, 372, 373, 374, 387

H

Haapane, Veija and Pettay study 709
 Hard and soft palate 364
 Hard palate 622
 Hawley appliance 162
 Hawley retainer 167, 180, 187
 Health care 781
 – financing and policy 780
 – history 691
 – worker 705
 Health Insurance and Accountability Act see HIPAA
 Hearing
 – acuity 607
 – deficiency 713
 – loss 355
 Hemifacial microsomia 286, 555–561
 Hemoglobin 317
 Hepatocyte, growth factor 309
 High density barium 617
 HIPAA (Health Insurance and Accountability Act) 694
 Holtz treatment protocol 390
 Horizontal lip tape 455
 Horizontal maxillary osteotomy 549
 Hospital admission 454
 Hospital for Sick Children 608
 Hotz and Gnoinski treatment principles 399
 Hotz presurgical orthopedic protocol 363, 364
 HOX gene 4, 5, 288
 HOXA2 9
 Huddart's passive orthopedic appliance 384
 Human Development Index (HDI) 252
 Human Subject Research Review Committee 21
 Hyaluronic acid 451
 Hynes pharyngoplasty 671, 672
 Hyoid bone 218, 622
 Hypernasal resonance 698
 – postpalatoplasty 708
 Hypernasal speech 648, 649, 652
 Hypernasality 52, 53, 327, 412, 418, 610, 618, 624, 629, 636, 649–651, 667, 669, 670, 676–678, 682, 705, 706, 708–711, 716, 735, 737, 739–741
 – postoperative 608
 Hypertonic lip musculature 117
 Hypertrophic tonsil 649
 Hypochondroplasia 290
 Hyponasal resonance 685
 Hyponasality 361, 618, 636, 649, 651, 672, 710
 Hypopharynx 653
 Hypoplastic adenoid tissue 647

Hypoplastic maxilla 535f, 538, 553, 564
 – fistula 535
 Hypoplastic velar muscles 510
 Hypotonic lip pressure 172
 Hyrax expander 142, 536

I

IBCLP (incomplete bilateral cleft lip and palate) 100, 110, 120, 143, 145, 165, 317, 318, 432, 238, 239, 470
 ICP (isolated cleft palate) 193ff, 225–227, 229, 230, 250, 251, 756
 – arch widths 249
 – chord lengths 249
 – overjets 249
 – tooth sizes 249
 – unoperated 249
 IDEA (Individuals with Disability Education Act) 706
 Ideal speech 487
 IDO (interdental distraction osteogenesis) 547, 548, 549
 IFN (Interferon)- γ 303, 309
 IL (Interleukin)-1 308
 IL-1 β 310
 IL-6 308, 309
 IL-10 308
 Iliac bone graft 277, 505
 Iliac crest 601
 Iliac crest bone graft 588, 598
 Ilium 463
 IMF see intermaxillary fixation
 Immunology 597
 Impedance testing 356
 Incisal canal 51
 Incomplete bilateral cleft lip and palate see IBCLP
 Incomplete unilateral cleft lip and palate see IUCLP
 Incomplete unilateral cleft lip see UICL
 Indian writings of Susrata 273
 Individual craniofacial structure 644
 Individual with facial deformities 263, 264, 265, 266, 268
 – with Disability Education Act see IDEA
 Infant orthopedics (IO) 397, 401, 402, 403, 404
 Inferior nasal conchae 626
 Infraorbital foramen 521
 Infraorbital neurovascular bundle 521
 Inheritance
 – autosomal dominant 204
 – X-linked 204
 Initial state 751, 752
 Innate language ability 607
 Insitute for Speech Disorders in Copenhagen and Arhus 225
 Inspiratory phase 662
 Institutional Review Board see IRB

Intact facial muscular force 43
 Intellectual capacity 607
 Intellectual development 259
 Intellectual functioning study 713
 Intelligence
 – verbal 258
 – nonverbal 258
 Intelligibility rating 403
 Inter-alveolar bony septum 604
 Interarch congruency 206
 Intercenter Comparison 768, 769
 Intercenter collaboration 756
 Interceptive orthopedics 489, 490, 491, 493, 495, 497
 – long-term prognosis 496
 Interdental distraction 551
 – osteogenesis (IDO) 535, 547–553
 – site 549
 Interdental osteotomy 549
 Interferon Regulatory Factor see IRF
 Interferon see IFN
 Interleukin see IL
 Intermaxillary fixation (IMF) 577
 Internal cortical flayer 556
 Internalizing behavior 713
 International Congress on Cleft Palate and Craniofacial Anomalies, Singapore 395
 International research strategy 775
 International Working Group 652, 653, 654
 Interplay of genetic and environmental factor 9
 Interpterygomaxillary disjunction 566
 Intracellular dye 3
 Intracranial dissection 568
 Intraoral flap 520
 Intraoral framework 480
 Intraoral maxillary protraction spring 538, 539, 547
 Intraoral molding plate 453
 Intraoral orthodontic splint 519
 Intraoral orthopedics 699
 Intraoral palatal expander 420
 Intraoral pressure 663, 664
 Intraoral splint 530
 Intraoral surgical manipulation 521
 Intraoral tooth-borne distraction device 545
 Intravelar veloplasty see IVV
 Intrinsic developmental deficiency 301
 Intrinsic growth deficiency 247, 304
 Iowa Pressure Articulation Test see IPAT
 IPAT (Iowa Pressure Articulation Test) 707
 IRB (Institutional Review Board) 694
 Island flap 329–331, 337
 – pushback 91, 326, 330, 333
 Isolated cleft palate see ICP
 IUCLP (incomplete unilateral cleft lip and palate) 48, 50
 IVV (intravelar veloplasty) 326, 461, 463, 685

J

Jaw discrepancy 583
 Jaw relation record 728
 Johansson Blizzard syndrome 530
 Juvenile diabetes 280

K

Kawamoto osteotomy 504, 510
 Kent Infant Development Scale 714
 Keratinocytes
 – allogeneic 307
 – autologous 307
 – biology 309
 – epidermal 307
 – growth factor 309
 – oral 307, 309
 – palatal 307
 Kernahan Rosenstein procedure 383, 397
 Kinesthetic feedback system 608
 Kirschner wire 176

L

Labial frenum attachment 453
 Labial osseous bridge 589
 Labial-palatal wire framework 480
 Labio-facial complex 750
 LAFH see Lower anterior facial height
 Laminin 4
 Lancaster Cleft Palate Clinic 277, 280, 735, 739
 Language
 – comprehension 258
 – development 699, 700, 712
 – stimulation 712
 Lateral cephalometric headplate 51
 Lateral cephalometric tracing 45, 117
 Lateral incisor 69, 70, 73, 92
 – pontic tooth 86
 Lateral pharyngeal wall 617, 637, 649, 653, 654
 Lateral port control 669, 670
 Lateral pterygoid plate 479
 Lateral skull X-ray 238, 239, 240
 Latham appliance 460, 463, 468, 469, 417, 423, 426
 Latham's pinned premaxillary retractive device 382
 Latham's presurgical orthopedic appliance 410, 414
 Latham-Millard pinned appliance 398
 Latham-Millard Presurgical Orthopedics Periosteoplasty with Lip Adhesion see POPLA
 Latham-Millard PSOT 434, 440, 443, 445
 Learning disability (LD) 713, 716

- Lefort I 108, 111, 144, 145, 147, 159, 206,
 331–333, 335, 444, 503, 506, 508, 512,
 514, 524, 525, 529, 530, 564, 573,
 575–580, 582, 583, 595, 609
 LeFort I advancement see LeFort I
 LeFort I maxillary advancement see
 LeFort I
 LeFort I osteotomy 503–507, 510, 512,
 514–518, 521, 547
 LeFort I surgery 418, 507, 608
 LeFort III 529, 564, 566, 568
 Length-for-age 401
 Levator veli palatini muscle 462, 625,
 644, 645
 LHX8 6p24 gene 9
 Lidocaine 618
 Lift/bulb prosthesis 740
 Linear changes 523
 Lingual collapse 590
 Lingual cortex 556
 Lingual crossbite 341
 Lingual tonsil 649
 Lip
 – adhesion 59, 68, 107, 317, 318, 319,
 320, 364, 398, 410, 411, 413, 417, 419,
 420, 454, 463, 588, 607
 – closure 279
 – revision 177
 – surgery 35, 315ff, 332, 334, 337, 411
 Listening tube 738
 Logan's bow 66, 109
 Lohmander-Agerskov study 698, 699
 Longitudinal cleft palate research study
 20
 Longitudinal dental cast study 21
 Longitudinal palatal and facial growth
 study 344
 Lower anterior facial height (LAFH) 65
 Lower posterior facial height (LPFH) 65
 LPFH see Lower posterior facial height
 L-shaped osteotomy 563
- M**
- MacArthur Communicative Develop-
 ment Inventory: Toddler 714
 Magnetic resonance imaging see MRI
 Major craniofacial conditions 777, 780,
 781, 782
 – history of care for children 778
 Major dysmorphia 463
 Major midface deficiency 463
 Major neurovascular bundle 306
 Malar-zygoma advancement 567
 Malignant hyperpyrexia 650
 Malnutrition 251, 252
 Malocclusion 111
 – class I 208
 – class II 105, 208, 320
 – class III 208, 210, 332, 333, 433, 444,
 463, 480, 503, 515, 516, 519, 563, 566, 573
 – residual 579
 Mammalian palate 8
 Mandible 21, 24, 622
 – morphology 30
 Mandibular arch 6, 43, 591
 Mandibular autorotation 160
 Mandibular bone stock 557
 Mandibular cast 721
 Mandibular central incisor 345
 Mandibular condylar cartilage 27
 Mandibular coordination 711
 Mandibular dental palatal cast 363
 Mandibular disproportion 582
 Mandibular distraction 555, 558, 561
 Mandibular downward-backward rota-
 tion 497
 Mandibular growth 23, 30, 64, 131, 145,
 218, 220–222, 233, 481
 Mandibular hypoplasia 286, 555, 557,
 569
 Mandibular micrognathia 52
 Mandibular osteotomy 409
 Mandibular overjet 341
 Mandibular plane angle 65
 Mandibular prognathism 110, 480, 496
 Mandibular prominence 340
 Mandibular pubertal growth spurt 123
 Mandibular ramus 32, 33
 – osteotomy 505
 Mandibular retrognathia 230, 496
 Mandibular sagittal split surgery 180
 Mandibular symposium 137
 Mandibulo-facial dysostosis 32, 635
 Masticating apparatus 721
 Masticatory muscle 561
 Matrix metalloproteinase see MMP
 – collapse 401
 Maxilla 21
 – anterior displacement 536
 – hypoplasia 590, 604
 – posterior displacement 536
 Maxillary alveolar bone 522
 Maxillary alveolar process 46
 Maxillary alveolar ridge 461
 Maxillary and mandibular arch congru-
 ency 448
 Maxillary anterior teeth 72, 96
 Maxillary anteroposterior osseous defi-
 ciency 589
 Maxillary arch 43, 51, 401
 Maxillary asymmetry 148
 Maxillary basal bone 336
 Maxillary bone deficiency 89
 Maxillary bone grafting 525
 Maxillary bridge 334
 Maxillary cast 721
 Maxillary central incision 186, 187
 Maxillary cleft defect 598
 Maxillary collapse 518
 Maxillary complex, shortening 317
 Maxillary constriction 662
 Maxillary cuspid 338, 480
 – area 491
 Maxillary deficiency 93, 506, 524, 525,
 528, 529, 530
 Maxillary dental midline 545
 Maxillary dental overjet 444
 Maxillary dental palatal cast 363
 Maxillary dentoalveolar retroclination
 63
 Maxillary dentoalveolus 557
 Maxillary distraction 525, 529, 564
 – osteogenesis 409, 411, 412, 519, 609
 Maxillary dysplasia 580
 Maxillary expansion 662
 Maxillary fistula 73
 Maxillary growth 23, 33, 64, 131, 304,
 583
 – deficiency 305, 487, 657
 – development 328
 – deviant 211
 – disturbance 160, 310
 – inhibition 279, 324, 326
 – vertical 391
 Maxillary hypoplasia 286, 479, 519,
 524, 526, 527, 530, 563, 567, 575–577,
 580, 750
 Maxillary midline 588
 Maxillary orthopedic protraction 547,
 548
 – effective 535, effective 538
 Maxillary osteotomy 119, 409, 662
 Maxillary prognathism 233, 497
 Maxillary prosthesis 278, 279, 729
 Maxillary protraction (Delaire facial
 mask) 111, 481, 493, 497, 535, 536
 – springs for effective maxillary ortho-
 pedic protraction 538
 – treatment protocol 538
 – treatment results 538
 – treatment effects 538
 Maxillary protrusion 64
 Maxillary relapse 508, 594
 – postoperative 506
 Maxillary retainer 438
 Maxillary retroclination 488
 Maxillary retrusion 151, 247, 480, 488
 Maxillary scarring 583
 Maxillary skeletal prognathism 495
 Maxillary surgical advancement 66
 Maxillary tuberosity 26, 220, 318, 349,
 462, 503, 505, 510, 536, 537
 Maxillary-mandibular relationship 63
 Maxillofacial animal research 598
 Maxillofacial growth 404, 697, 700
 Maxillo-facial skeletal complex 750
 Maxillofacial trauma reconstruction
 530
 Maxillo-mandibular distraction 559,
 560
 Maxillo-mandibular fixation 663
 Maxillo-mandibular growth 573
 Maxillo-mandibular skeletal discrep-
 ancancy 496
 Maxillo-mandibular surgery 111

- Maximum-maxillary sagittal advancement 529
 McComb procedure 463
 McComb suture 468, 469, 470
 McNeil and Burston technique 399
 McNeil's orthopedic procedure 381, 382
 MDI (Mental Development Index) 715
 Mean Length of Utterance 714
 Medial canthal ligament 566
 Medial epithelial edge (MEE) 8
 Medical autonomy 779
 – limits 779
 Medical decision-maker 779
 MEE see medial epithelial edge
 Meningeal disruption 568
 Mental Development Index see MDI
 Mental retardation 264
 Mesenchymal cell deficiency 8, 752
 Mesenchymal stem cell 597, 599
 Mesial molar occlusion 341
 Mesioangular rotation 71, 182, 207, 345, 481
 Mesoderm 288
 Meta-analysis of the existing literature 404
 Method of total nose reconstruction 273
 Miami Children's Hospital 459
 Miami Craniofacial Anomalies Foundation 365, 409
 Microglossia 218
 Micrognathia 215, 218, 221, 222, 227, 555, 561, 562, 569
 Micrognathic mandible 215, 216, 218
 Microstomia 361
 Middle frontal gyrus (MFG) 711
 Middle-ear
 – effusion 355, 356, 359, 360, 361
 – pressure 360
 Midface deficiency 526, 563
 Midface elongation-rotation movement 561
 Midface hypoplasia 567, 568, 577
 Midface osteotomy advancement 570, 571
 Midface retrusion 567, 569
 Midface's forward growth 446
 Midfacial deficiency 411
 Midfacial growth 137, 172, 305, 326, 329, 375, 409, 417, 479, 484, 488
 – inhibition 194, 317, 397
 – retardation 336, 340
 – restraining forces 109
 Midfacial hypoplasia 512, 635
 Midfacial orthopedic protraction 479
 Midfacial osteogenesis 415
 Midfacial protrusion 36, 90, 131, 141, 340, 484, 760
 Midfacial recession 208
 Midfacial recessiveness 424
 Midfacial retrusion 112, 381, 418, 479, 480, 487, 491, 508
 Midline deviation 341
 Midsagittal plane 452, 453, 616, 625
 Millard approach 463
 Millard forked flap 111, 113, 114, 115, 132
 Millard Island flap 640
 – pushback procedure 329
 Millard lip closure 491
 Millard periosteoplasty 279
 Millard protocol 460
 Millard rotation advancement 68, 79, 275, 345, 387
 Millard surgical lip procedure 112
 Millard technique 401
 Millard-Latham approach 460
 Millard-Latham method 398
 Millard-Latham PSOT see Latham-Millard PSOT
 Minimal morbidity 581
 Minnesota Child Development Inventory 714
 Minority group 265
 Minority population 781
 Misarticulation 708
 Mismatch negativity see MMN
 Mitochondrial abnormality 657
 Mixed oral-nasal breather 658
 MMN (Mismatch negativity) 716
 MMP (Matrix metalloproteinase) 303, 304
 MOLDING 319
 Molecular biology 597
 Molecular craniofacial biology 3
 Monoblock osteotomy 564, 568
 Monoclonal antibody 3
 Mouth breathing 658, 660, 685
 MRI (magnetic resonance imaging) 669, 695
 MSX1 9
 Mucoperiosteal flap 277, 304, 323, 324, 327, 328, 349, 461, 601, 605
 Mucoperiosteal tissue 194
 Mucoperiosteal undermining 320
 Mucoperiosteum 65, 67
 – insufficient 119
 Mucosal biopsy 307
 Mucosal closure of the alveolus an anterior hard palate cleft 277
 Mucosal epithelium 357
 Mucosal recess 489
 Mucosal split flap 306
 Mucosal tolerance to ulceration 454
 Multicenter cephalometric study 385
 Multicenter study 598
 Multidisciplinary surgical and research program 237
 Multidisciplinary approach 732
 Multidisciplinary team 396, 459, 685
 Multifactorial inheritance 9
 Multi-institution involvement 268
 Multiple sclerosis 637
 Multiview fluoroscopy 683
 Multiview videofluoroscopy 615, 616, 617, 667, 677, 651, 655, 654
 – advantages 654
 – disadvantages 654
 Murphy's law 654
 Muscle constriction 746
 Muscle fiber hypoplasia 657
 Muscle tonicity 608
 Mutual understanding 719
 Myasthenia gravis 650, 735, 740
 Myelomeningocele 280
 Myofunctional therapy 746
 Myringotomy 361
- N**
- Nagers's syndrome 555
 NAM (nasolabial molding) 390, 451–453, 457, 458, 460, 461, 463
 – bilateral 456
 Narrow shelves 753
 NASA (National Aeronautics and Space Administration) 755
 Nasal acoustic energy 618
 Nasal airflow 619
 – disorder 706
 Nasal airway 657, 658, 659, 660, 661, 662
 – effects on speech 663
 – obstruction 667
 – resistance 488, 657, 658, 662, 663, 664
 Nasal alar base 453
 Nasal alar cartilage 46
 Nasal Alveolar Molding see NAM
 Nasal breather 658, 660, 662, 663
 Nasal capsule deficiency 657
 Nasal congestion 651
 Nasal dorsum 472
 Nasal endoscopy 721, 738, 741
 Nasal mucoperiosteum 324, 510
 Nasal mucosa 593
 – flap 674
 – lining 456
 Nasal pharynx 730
 Nasal polyps 662
 Nasal resonance 643, 722
 Nasal septal cartilage 27
 Nasal septum 62, 80, 83, 211, 384, 396, 397, 451, 625
 – growth 103
 – hypertrophy 657
 – theory 25, 27
 Nasal skeleton 27
 Nasal soft tissue floor 589
 Nasal stent
 – unilateral 453, 454
 – hard acrylic 455
 Nasal surgical procedure 400
 Nasal symmetry 605
 Nasal tip cartilage 276
 Nasal turbulence 682
 Nasal valve 659, 660
 Nasal velar mucosa 674

Nasal-lip revision 111
 Nasendoscopy 610, 667, 683, 685
 Nasoalveolar molding 395
 – Grayson 399
 Nasoalveolar molding see NAM
 Nasoendoscopic guided clinical examination 581
 Nasoendotracheal anesthesia 549
 Nasolabial angle 391, 570
 Nasolabial esthetics 452, 457
 Nasolabial outcome 769
 Nasolacrimal groove 566
 Nasomaxillary complex 24, 25, 104
 Nasometer 651
 Nasopalatine foramen 51, 193, 218, 220
 Nasopharyngeal airspace 661
 Nasopharyngeal airway 662
 – obstruction 673, 675
 Nasopharyngeal area 21
 Nasopharyngeal growth 622, 623
 Nasopharyngeal skeletal architecture 621ff
 Nasopharyngeal sphincter 640
 Nasopharyngeal structure 731
 Nasopharyngoscopy biofeedback 644
 Nasopharyngoscopy 462, 615, 617, 624, 636, 653, 654, 655, 697
 – advantages 654
 Nasopharynx 53, 621–623, 628, 632, 637, 639, 653, 682
 National Aeronautics and Space Administration see NASA
 National Institute of Dental Research (NIDR) 404
 National policies 771
 Negative feedback 265
 Negative stereotype 263
 Nemaline myopathy 650
 Neonatal hypoxia 650
 Neonatal infant orthopedics 400
 Neonatal intensive care 777
 – unit see NICU
 Neonatal maxillary orthopedic treatment 26
 Neonatal maxillary orthopedics 110, 112
 – history 395ff
 – intraoral 110, 342, 381ff, 751,
 Neonatal orthopedics 401
 Netherlands Approach 385
 Neural crest 288
 Neurodegenerative disease 650
 Neurofibromatosis (von Recklinghausen's disease) 5
 Neurogenic deficiency 726
 Neurologic disorder 626, 637, 650, 735
 Neuromuscular deficit 732
 Neuromuscular disorder 667
 Neuromuscular bundle 556
 Neurovascular damage 558
 Nice dental smile 497
 β -Nickel-titanium helix spring 538
 NICU (Neonatal intensive care unit) 778, 779

NIDR see National Institute of Dental Research
 Nijmegen Cleft Palate Center in The Netherlands 762
 Non-cleft craniofacial abnormalities 291
 Non-minority population 781
 Nonphysiological surgery 327
 Non-POPLA (Nonpresurgical orthopedics) 409, 414, 417, 444, 446, 447, 448
 Nonprothodontic dental rehabilitation 601
 Nonresorbable polymer membrane 307
 Normal anatomy 396
 Normal palate 376
 Normal speech 691
 Northwestern University Dental School 325
 Nostril apex 455,
 – overhanging 451, 452
 Nostril occlusion 651
 Nostril stenosis 461
 Nurse Coordinator 459
 Nursing 291
 – prenatal 291
 – neonatal 291
 – infant/toddler 291
 – preschool/school-aged/adolescents 291
 Nutrition 286

O

Obstructive sleep apnea see OSA
 Obturator 719, 720
 Occlusal disaster 557, 561
 Occlusion 201, 204
 – class I 126, 147, 150, 169, 194, 210, 418, 595
 – class II 79, 82, 126, 134, 135, 147, 210, 344, 376, 418
 – class III 344, 376
 Oculopharyngeal muscular dystrophy 650
 Orthodontic discipline 396
 Operating microscope 355, 356
 Optical profilometer 755
 Oral airway impedance 662
 Oral and maxillofacial surgery 292
 – adolescents 292
 – infant 292
 – neonatal 292
 – prenatal 292
 – preschool 292
 – school-aged 292
 – toddler 292
 Oral breather 658, 660
 Oral cleft 13, 14
 – prenatal diagnosis 13, 15
 Oral commissure 557
 Oral Facial Evaluation 512
 Oral manometer 738
 Oral motor coordination 607
 Oral mucosa closure 323
 Oral nasal air pressure 741
 Oral velar mucosa 674
 Oral-facial handicap 721
 Oral-nasal balance 709
 Ora-nasal breather 662
 Orbicularis muscle 273
 – fiber 276
 Orbicularis oris 43, 44
 – muscle 400, 510
 Orbicularis-oris-buccinator-constrictor pharyngis superioris muscle ring 753
 Orbit 21
 Orbital hypertelorism 287
 Orofacial cleft 8, 519
 Orofacial clefting genes 774
 Orofacial digital syndrome 203
 Orofacial function 488
 Orofacial growth 376, 390
 Oronasal fistula 457, 489, 575, 577, 578, 590, 596, 605, 675
 – closure 575, 579, 580
 – recalcitrant 577
 – repair 576
 Oronasal opening 77
 Oropharyngeal membrane 5
 Oropharynx 53, 622, 623, 653
 Orthodontic appliance 724, 725
 Orthodontic arch wire 85
 Orthodontic closure 604
 Orthodontic elastic band 453, 454
 Orthodontic expansion 66
 Orthodontic management 286
 Orthodontic stimulation 591
 Orthodontic strategy 487
 Orthodontic surgical treatment 119
 Orthodontic tooth eruption 587, 588
 Orthodontic treatment 82, 89, 92, 155, 201, 204, 280, 325, 367, 565
 – final 82
 – method 316
 – presurgical 35
 – standard 753
 – surgical 20
 Orthodontic-orthopedic forces 91
 Orthodontics 292
 – adolescents 293
 – adults 293
 – infant 292
 – neonatal 292
 – preschool 292
 – school-aged 293
 – toddler 292
 Orthodontist 459, 573
 Orthognathic surgeon 608
 – preoperative considerations 610
 Orthognathic surgery 286, 460, 497, 530, 573, 581, 587, 590, 595, 607ff
 – speech implication 607ff

Orthognathic-orthodontic procedure 173
 Orthopedic force application 599
 Orthopedic plate 327
 Orthopedic protraction forces 60
 Orthopedic treatment, early 396
 Orthopedic-orthodontic CLP treatment 488, 489
 Orthopedic-orthodontic appliance system 487
 Orticochea 640
 Orticochea pharyngoplasty 673, 707
 OSA (obstructive sleep apnea) 641, 650, 667, 670, 671, 673, 685
 Oslo CP Center 491
 Oslo CLP team 108, 351
 Oslo study 64, 109
 Osseous CP patient 600
 – rehabilitation 600
 – restoration 600
 Osseous maturation 587
 Osseous particulate autogenous graft 599
 Ossicular chain 361
 Osteoblastic activity 23
 Osteoclastic-osteoblastic remodeling 596
 Osteogenic appliance 412
 Osteogenic deficiency 26, 90, 91, 156, 198, 328, 342, 375, 750
 Osteolysis 543
 Osteosynthesis 512
 Otitis media 355, 361
 – acute 357
 – with effusion 357
 Otolaryngologic examination 657, 658
 Otolaryngologist 459, 573, 654, 716
 Otolaryngology 293, 683
 – adolescent and adult 294
 – neonatal 293
 – prenatal 293
 – preschool 294
 – school-aged 294
 – toddler 294
 Otorrhea 355
 OTX gene 288
 Overall strategy 459
 Overbite/overjet relationship 134, 135, 149, 150, 155, 158, 165, 173, 346, 437, 448, 506
 Overt cleft 646

P

Palatal acrylic 386
 Palatal aponeurosis 674
 Palatal arch
 – expansion 331
 – form 43ff
 – symmetrie 381
 – wire 495
 Palatal bone graft 504

Palatal cast 56
 Palatal Cast Study 762
 Palatal cast's surface 363
 Palatal cleft closure 68, 81, 88, 94, 114, 116, 326, 343, 344, 375
 – procedure 364
 Palatal efficiency rating computed see PERCI
 Palatal embryopathology 752
 Palatal expander 71, 346
 Palatal expansion 88, 203, 211
 Palatal fistula 74, 459, 462, 524, 593
 Palatal flap 592, 601
 Palatal fusion 52
 Palatal growth 21, 23, 25, 75, 79, 84, 127, 193, 219, 326, 377
 – acceleration 138, 139
 – comparison 363
 – posterior 366
 – study 749, 752
 Palatal helix expander 204, 208
 Palatal hypoplasia 102
 Palatal insufficiency 53, 627
 Palatal left closure 169
 Palatal lift appliance see PLA
 Palatal lift prosthesis 735, 738, 739, 741, 742, 743, 744
 Palatal maldevelopment 416
 Palatal mucoperiosteum 302, 315, 343, 640
 Palatal mucosa 302, 308, 325, 719
 Palatal osteogenesis 449
 Palatal osteogenic deficiency 113, 448, 669
 Palatal osteotomy 336
 Palatal point 722
 Palatal retainer 69, 183
 Palatal scar, 519
 – contracture 207
 – scar tissue 306
 Palatal segment's collapse 207
 Palatal shelves 63
 – size and shape 63
 Palatal surgery (s. also palate surgery) 310, 460, 762
 – conservative 327, 328
 – nontraumatic 328
 – timing 696
 Palatal surgical closure 366
 Palatal vault space 133
 Palatal wound healing (s. also wound healing) 301
 Palate cleft closure 328
 Palate closure 66, 463
 Palate
 – primary 285
 – secondary 285
 Palate growth 66
 Palate repair
 – delayed complete 328
 – early complete 328
 – early lip and soft 328
 – late complete 328
 Palate surgery (s. also palatal surgery) 315ff, 332, 334
 Palate's arch form 35
 Palates mucoperiosteum 21
 Palatine artery 593
 Palatine foramen 324
 Palatine tonsil 649
 Palatine vessel 462
 Palatogenesis 8
 Palatoglossus 624
 Palatopharyngeal closure 638
 Palatopharyngeal fold 622
 Palatopharyngeal incompetence 639
 Palatopharyngeal insufficiency 741, 743, 744
 Palatopharyngeal valve 719
 Palatopharyngeus 640
 Palatoplasty 63, 64, 107, 329
 – and speech outcome 694
 – technique 695
 – timing 693
 Panorex 173, 175
 – postoperative 174
 – preoperative 173
 Parental adjustment 259
 Parental decision 780
 Parental responsibility 777
 Parental stimulation 695
 Parent-infant bonding 695
 Parents' satisfaction 400, 401
 Parkinsonism 637
 Particulate marrow and cancellous bone see PMCB
 Passavant's pad 624
 Passavant's ridge 644, 645
 Pathological adjustment 266
 Patient dissatisfaction 769
 Patient noncompliance 591
 Patient's phenotype 326
 PAX gene 289
 PDGF (platelet-derived growth factor) 302
 Pediatric dentistry 294
 – adolescent 294
 – adult 294
 – infant 294
 – neonatal 294
 – prenatal 294
 – preschool 294
 – school-aged 294
 – toddler 294
 Pedodontist 459
 Peditrician 716
 Pennsylvania State Palate Program 277
 PERCI (palatal efficiency rating computed) 619
 Perialar musculature 659, 660
 Periodontal deficiency 592
 Periodontal fiber system 310
 Periodontium 605
 Periosteoplasty 107, 391, 398, 416, 419, 423, 432, 434, 491, 495, 588
 Peristalsis 623

- Permanent dental injury 558
 Permanent dentition 60, 317, 320
 Pernalisity 510
 Perpendicular plates of the sphenoid 344
 Pfeiffer syndrome 287, 530
 Pharyngeal airway 216, 217, 649
 Pharyngeal arch 6–8, 288, 289
 Pharyngeal bulb 638, 639
 Pharyngeal extension 193
 Pharyngeal flap 53, 181, 459, 462, 503, 609, 610, 617, 635–637, 661, 670, 684, 686, 719, 723
 Pharyngeal flap surgery 650, 707, 737, 738, 741, 742
 – complications 671
 Pharyngeal flap technique 320
 Pharyngeal grooves 289
 Pharyngeal hypotonia 641, 647
 Pharyngeal implant 641
 Pharyngeal mucosa 670
 Pharyngeal pouches 288, 289
 Pharyngeal scarring 519
 Pharyngeal section 729, 731
 Pharyngeal skeletal architecture 52
 Pharyngoplasty 507, 669, 705, 708, 732
 – primary 710
 – secondary 710
 Pharynx 621–623
 Phenylephrine 618
 Philtral column 275, 276
 Physical attractiveness stereotype 264
 Physical therapy 739
 Physical therapy modality 726
 Physiological surgical procedure 363, 377
 Pierre Robin sequence 215ff, 286, 328, 361, 555, 561, 667, 671, 673, 750
 PLA (palatal lift appliance) 639, 640
 Plastic surgeon 459, 735
 Plastic surgery 294
 – adolescent 295
 – adult 295
 – infant 295
 – neonatal 295
 – prenatal 295
 – preschool 295
 – school-aged 295
 – toddler 295
 Plastic surgery 491, 683
 Platal cleft-nasal floor defect 504
 Plate-and-screw fixation 577
 Platelet-derived growth factor see PDGF
 Platybasia 647
 PMCB (particulate marrow and cancellous bone) grafting 587–591, 595, 598
 Pneumatic otoscopy 355
 PNS (posterior nasal spine) 522, 536, 722
 Polio 735
 Polysomnogram 675
 Polysomnographic evaluation 671
 Polythelia 203
 POPLA (presurgical orthopedics and periosteoplasty with lip adhesion) 409, 410, 411, 414–418, 426, 428, 430, 431, 433, 446–448
 Popliteal pterygia 203
 Posnick procedure 448
 Postdistraction 523, 524, 526–528
 – alveolar bone grafting 551
 – orthodontic tooth movement 550, 551
 Posterior cleft space 130, 136, 137, 156, 157, 170, 171, 185, 370, 373
 Posterior crossbite 489
 Posterior displacement of maxilla 536
 Posterior nasal spine see PNS
 Posterior pharyngeal flap 667–671; 673, 677
 Posterior pharyngeal wall 627, 629, 632, 633; 635, 638; 653; 676
 – augmentation 641, 675
 Posterior rotation of the mandible 392
 Posterior teeth splint 78
 Posterior wall of the pharynx 243, 244
 Postmaxillary distraction osteogenesis 413
 Postnatal mandibular growth spurt 484
 Postobstructive sleep apnea 682
 Postoperative airway obstruction 670
 Postoperative nasendoscopy 673
 Postoperative orthodontic treatment 515
 Postpharyngeal flap nasendoscopy 686
 Postpubertal facial growth period 336
 Postpubertal facial growth spurt 158
 Postsurgical psychosocial therapy 417
 Posture of the tongue, abnormal 222
 Preauricular incision 563
 Predistraction 523, 524, 526–528
 Prejudice against individuals with clefts 263
 Prelexical development 699
 Premaxilla protrusion 109–113, 132, 229, 230, 233, 389, 410, 413, 414, 416, 428, 430, 433, 469, 484
 Premaxilla relapse 595
 Premaxilla retrusion 411–413
 Premaxilla ventroflexion 126, 137–139, 151, 179, 182, 429
 Premaxilla's palatal incline 206
 Premaxillary deformity 541
 Premaxillary excision 178
 Premaxillary growth 170, 171
 Premaxillary incision 177
 Premaxillary orthopedic intrusion 535, 542
 – device 541
 – mechanisms 543
 – orthodontic preparation 541
 – treatment results 541
 Premaxillary orthopedic medial repositioning 535, 544, 546
 Premaxillary orthopedic medial repositioning, device 545
 – mechanisms 545
 – treatment results 545
 Premaxillary overbite 109, 187, 188, 321, 322
 Premaxillary overjet 188, 321, 322
 Premaxillary protrusion 36, 45–47, 50, 99, 100, 103, 104, 107, 108, 115, 117, 119, 143, 145, 148, 151, 154, 158, 186, 760
 Premaxillary setback 107, 108
 Premaxillary surgical setback 116, 118, 126, 128, 175, 179
 Premaxillary ventroflexion 37, 320
 Premaxillary vomerine suture see also PVS 44, 45, 49
 Premaxillary-vomerine complex 229, 230
 Prenatal diagnosis 778, 780, 782
 Prenatal maxillary hypoplasia 487
 Prenatal screening mechanisms 780
 Prepalatal cleft 273
 Prepubertal growth spurt 118
 Preschool investigation 714
 Presurgical maxillary orthopedics 328
 Presurgical orthodontic treatment 35
 Presurgical orthodontics 334
 Presurgical orthopedic treatment (s. also PSOT) 277
 Presurgical orthopedics 278, 343, 364, 367, 375, 382, 383, 395, 396, 446, 490, 749, 752
 Presurgical orthopedics and periosteoplasty with lip adhesion see POPLA
 Presurgical palatal manipulation 381
 Presurgical treatment 460
 Primary bone graft 278, 279, 382–384, 391, 397
 Primary cleft surgery 774
 Primary hypoplasia 247
 Primary nasal correction 461
 Primary surgical cleft repair 458
 Primary surgical closure 342
 Primary veloplasty 696, 697
 Programmed cell death 5–7, 9
 Prolabial band 456
 Prolabial vermilion 461
 Prolabium 47
 Prominent premaxilla 351
 Prospective study 20
 Prosthetic appliance 77
 – anterior 78
 – removable 77
 Prosthetic dental appliance 637
 Prosthetic dentistry 732
 Prosthetic rehabilitation 720
 Prosthetic speech aid 724
 Prosthetic speech appliance 719–721, 723, 725–729
 Prosthetic stimulation 745
 Prosthetic treatment 732
 Prosthodontic reconstruction 598

Prosthodontic treatment 280, 599
 Prosthodontist 719, 723, 732, 735
 Protein
 – assay 3
 – cytoskeletal 309
 Protraction facial mask 66, 425, 426, 433, 438, 479, 482, 487, 493
 – treatment results 495
 Protraction orthopedics 161
 PSOT (presurgical orthopedic treatment) 38, 39, 99, 343, 376, 382, 384–387, 389, 391, 396, 397, 399, 400, 414, 415, 416, 418, 436, 440
 – clinics 390
 Psychoeducational development 692
 Psychological adjustment 257, 721, 264, 267, 268
 Psychology and clinical social work 296
 – adolescent 297
 – adulthood 297
 – infant 296
 – neonatal 296
 – prenatal 296
 – preschool 296
 – school-aged 296
 – toddler 296
 Psychopathology 264
 Pterygoid plate 25, 632
 – of the sphen 503
 Pterygomaxillary fissure 349, 364
 Pterygomaxillary gap 504, 510, 512
 Pterygo-maxillary junction 561, 564
 Pterygomaxillary space 510
 Pterygomaxillary suture see PTN
 Pterygomaxillary tuberosity 568
 PTN (pterygomaxillary suture) 315
 Ptosis of the tongue 216
 Pulmonic stenosis 14
 Pure-tone testing 356
 Pushback procedure 319, 374, 640
 Pushing effect 315
 PVRL1 9
 PVS (premaxillary vomerine suture) 99, 100, 102–104, 110, 113, 115, 172, 188, 315, 316, 410, 413, 414, 416, 429, 432, 446–484

Q

Quad-helix fixed appliance 564
 Quad-helix spring 491, 494, 495
 Quantitative palatal parameter 343

R

Radiographic evaluation 741
 Randomized clinical trial (RCT) 20
 Rapid maxillary expander 547
 Rapid maxillary expansion (RME) 535, 538, 540

Reading disability 713
 Reasoning ability 258
 Reconstructive surgery 305
 Reference group 268
 Reflex microscope 401
 Reflex microscopic analysis 238
 Regression analysis 707
 Removable maxillary prosthesis 156
 Residual bony defect 596
 Residual deformity 268, 575
 Resilient child 257
 Resonance balance 639
 Resource allocation 780
 Respiratory classification 657
 Respiratory effort 663
 Respiratory embarrassment 218
 Retinoic acid 7
 Retrognathia 64
 Retrognathic mandible 115
 Retrognathic profile 222
 Retrospective study 20, 21
 Revascularization 604
 Reversible growth hormone resistance 251
 Rhesus monkey 360
 Rhinoplasty 274, 510, 779
 Rhombomere 289
 Rigid external distraction 522, 524, 529, 530
 – system 520
 RME (rapid maxillary expansion) 538, 540
 Robin sequence 5, 226, 227, 230
 Roentgencephalometry 21
 Root absorption 153
 Rosenstein appliance 397
 Ross's multicenter study 64, 65
 Rotation advancement lip surgery 434
 Rotation advancement technique see Millard's rotation advancement
 Round house bridge 335
 Routine Clinical Audit 769
 Rowe forceps 510, 558
 Rowe Kiley forceps 566, 569
 RS 608
 Rush Medical College, Chicago 276

S

Saethre-Chotzen syndrome 287
 Sagittal jaw discrepancy 496
 Sagittal jaw relation 64
 Sagittal maxillary relapse 525
 Sagittal ramus osteotomy 575, 578
 Sagittal splitting procedure 505
 SAS Proc Mixed software 365
 Scandcleft study 769
 Scar formation 301
 School achievement 258, 259, 713
 School-aged investigation 714
 Schuchardt procedure 515
 Scientific evidence 692
 Scott's hypothesis 25, 26, 27, 398
 Secondary bone grafting 603, 604
 Secondary bone necrosis 568
 Secondary epithelialization 516
 Secondary midfacial malformation 495
 Self-acceptance 264
 Self concept 264
 Self-esteem 263, 264, 265, 266, 267, 268
 Self-fulfilling prophecy 264
 Self-protective property 264–267
 Self-protective strategy 265, 267, 268
 Sella turcia 18, 522
 Sella-nasion line 17
 Septal deviation 212
 Septoplasty 575, 578
 Septo-premaxillary ligament 104
 Serial cephalometric tracing 341
 Serial cephaloradiograph 17
 Serial facial growth study 752
 Sharpey's fiber 25, 305–307
 Silicone augmentation pharyngoplasty 676
 Skeletal angle of convexity 529
 Skeletal cephalometric landmark 232
 Skeletal deformity, secondary 324
 Skeletal growth 23
 Skeleto-muscular deficiency 753
 Skin expansion 561
 Skin incision 276
 Skoog's periosteal flap 278
 Skull 21
 – base 358
 Sleep apnea (s. also OSA) 519
 Small midline gap 685
 Social adjustment 259, 264, 266
 Social and emotional functioning 257
 Social comparison 266
 Social competence 267
 Social discrimination 265
 Social interaction feedback system 264
 Social justice 780
 Social psychological research on facial deformity 268
 Social rejection 268
 Social stigma see stigma
 Socio-economic background 691
 Socio-economic status 268
 Soft palate 55, 60
 Soft palate pushback procedure 334
 Soft tissue cleft 60
 Soft-tissue contraction 324
 Soft tissue matrix 25
 Soft tissue maxillary retrusion 64
 Sonic hedgehog 289
 Sonic hedgehog (Shh) gene 4
 Sound spectrographic analysis 721
 South Florida Cleft Palate Clinic 365, 398, 409, 414
 SP (sphincteric pharyngoplasty) 640, 641
 Spatial relation 724

- Special Research Fellowship from the National Institute of Dental Research Institutes of Health 222
 Speech acceptability 615, 721
 Speech acquisition 644
 Speech aerodynamics 664
 Speech aid appliance 637, 638
 Speech aid prosthesis 661, 662
 – contraindication 726
 Speech appliance 719
 Speech articulatory disorder 741, 746
 Speech and language development 400, 403, 696, 700
 – evaluation 402
 Speech and language pathology 297
 – adult 298
 – neonatal and infancy 297
 – preschool 298
 – school-aged 298
 – toddler 297
 Speech bulb 729, 731, 732
 Speech characteristics 740
 Speech deficiency 713
 Speech development 607, 750
 Speech disorder 252, 626
 Speech examination 707
 Speech habilitation 639
 Speech intelligibility 615, 650, 678, 684, 699, 739
 Speech-language evaluation 682
 Speech-language pathologist 325, 326, 375, 376, 448, 573, 581, 607, 608, 610, 615, 616, 635, 637, 654, 683, 692, 700, 705, 712, 716, 717, 735, 739
 Speech learning 705
 Speech mechanism 610, 696
 Speech occlusion 326
 Speech performance 663
 Speech physiologist 616
 Speech production 616, 649
 Speech proficiency 619, 707, 708
 Speech readiness period 705
 Speech recordings 721
 Speech sound acquisition 700, 709
 Speech testing procedure 741
 Speech therapist 459
 Speech therapy 280, 376, 403, 618, 636, 640, 698, 699, 711, 712, 717, 740, 746
 – role of parents 712
 Spheno-ethmoidal suture 29
 Spheno-ethmoidal/circumaxillary suture system 28
 Spheno-occipital synchondrosis 28, 623
 Sphincter pharyngoplasty 667, 668, 671–673, 677, 678, 684, 685, 710
 – surgery 618
 Sphincteric muscle system 319
 Sphincteric pharyngoplasty see SP
 Splanchnocranial skeleton 27
 Split flap technique 310
 Square-shaped flap 275
 Sri Lanka Human Development Index 252
 Sri Lankan Cleft Lip and Palate Project 237, 251, 252
 Standardized speech sample 652
 Steel crushing clamp 276
 Steering Group 771
 Steinert syndrome 650
 Stereophotogrammetry 140
 Stereophotogrammetric electromechanical 3D digital analysis 22
 Stertorous breathing 220
 Stigma 264
 Stigmatized group 263–268
 Stigmatized individual 266
 Stigmatizing condition 268
 Stimulation of bone growth 26
 Stimulator pad 381
 Stroke 735
 Structural disorder 649
 Subglottal pressure 664
 Submucous cleft 50
 Submucous cleft palate 199ff, 648
 – occult 648, 649
 Subperiosteal dissection 564
 Subperiosteal onlay rib graft 397
 Sucking 711
 Superior based pharyngeal flap 66
 Superior constrictor 43, 44
 Superior pharyngeal constrictor muscle 670
 Superior temporal gyrus (STG) 711
 Superiosteal dissection 503
 Supramarginal gyrus 711
 Supraorbital rim 566
 Surgical denervation 659
 Surgical distraction osteogenesis 553
 Surgical iatrogenesis 230, 233
 Surgical maxillary advancement 514
 Surgical morbidity 454, 522
 Surgical outcome, prognosis 452
 Surgical planning 669
 Surgical protocol 343
 Surgical reinnervation 659
 Surgical trauma 305
 Surgical treatment 316
 – method 316
 – planning 363
 – staged 319
 Surgical-orthodontic treatment 20, 111
 Surrounding palatal tissue 363
 Sutural contraction osteogenesis 543
 Sutural push theory 25
 Suture-brachycephaly, bicoronal 287
 Suture-plagiocephaly, unicoronal 287
 Suture-scapocephaly, sagittal 287
 Suture-trigonocephaly, metopic 287
 Swallowing 623, 625, 722, 729–732
 – 22q11 syndrome 280
 Synostosis 103
- T**
 Tang Dynasty 273
 Team evaluation 285
 Teeth
 – development 288
 – malformed 203, 208
 – missing 203, 208
 – splinting 205
 – supernumerary 208
 Tegaderm 457
 Templin-Darley Screening test of Articulation 707
 Temporal facial graft 463
 Temporalis muscle flap 462
 Temporomandibular joint 32
 Temporomandibular joint
 – ankylosis 555, 563
 Tennison procedure 226
 Tensor veili palatini 357
 – muscle 624
 Teratogen 8
 Tetracain hydrochloride 618
 TFH see total facial height
 TGF (transforming growth factor) 302
 TGFA 9
 TGFB3 9
 TGFβ 303, 304, 309
 Thanatophoric dysplasia 290
 Three-dimensional measuring technique 754, 755
 Tibia 463
 Tibial periosteum 278
 Timing of surgery 582
 Timing of treatment 459
 Tissue engineering 306, 308, 597
 Tissue irritation 456
 Tissue regeneration 307
 Tissue shortage 317
 TONAR (The Oral-Nasal Acoustic Ratio) 619
 Tongue resistance 753
 Tonsillar hypertrophy 671
 Tonsillar size 668
 Tonsillectomy 52, 650, 684, 669
 Tooth eruption 76
 Toronto Hospital for Sick Children 280
 Total facial height (TFH) 65
 Total maxillary advancement 510
 Total surface area 368, 370, 373
 Towne's view 616, 617, 653
 Tracheotomy 216, 218, 221
 Traditional speech treatment 740
 Transforming growth factor see TGF
 Transitional dentition stage 530
 Transpalatal scar contracture 338
 Transverse expansion 493
 Transverse maxillary deficiency 582
 Transverse maxillary osteotomy 521, 529
 Transverse maxillary widening 493
 Traumatic brain injury 740
 Traumatic injury 739

Tramatic surgery 381
 – on midfacial growth 325
 Treacher Collins syndrome (TCS) 5,
 227, 286, 287, 361, 561, 775
 Treatment plan 351
 Treatment planning 344, 376
 Tretrospective clinical trial 21
 Triangular flap 275
 Trisomy 13 14
 Trisomy 18 14
 Trunk neural crest 4
 Tubal dysfunction 359, 360
 Tubal evaluation 359
 Tubal obstruction 359
 Turbinate hypertrophy 662
 Two-layer palato-vomeroplasty 341
 Tympanic membrane 355, 357, 359–361
 Tympanometry 356, 359
 Tympanosclerosis 361
 Tympanum 6

U

UAFH see upper anterior facial height
 UCLP (unilateral cleft of the lip and
 palate) 26, 63, 65, 207, 210, 226–233,
 237–239, 245, 252, 327, 329, 385,
 397–401, 403, 487, 495–497, 575, 578,
 579, 664, 693, 696, 708, 753, 772, 774
 – nonsyndromic 404
 – sequence of operations for the repair
 772
 – unoperated 253, 254
 UCLP Study Models Analysis 245–247
 – arch widths 245, 247
 – chord lengths 246
 – crossbites 246
 – crowding 246
 – missing teeth 246
 – overjet 246, 247
 – tooth widths 245
 UICL (incomplete unilateral cleft lip)
 227, 229–233
 Ultrasound
 – classification 13
 – three-dimensional 14
 – transvaginal 14
 Unilateral alveolar gap 454
 Unilateral cleft lip and palate 521
 – patients 538
 Unilateral cleft lip nose 273
 Unilateral cleft of the lip and palate see
 UCLP
 United Nations Development Program
 252
 University of Miami School of Medicine
 409
 Upper anterior facial height (UAFH) 65
 Upper labial sulcus 517
 Upper lip protrusion 132
 Uveopalatoplasty 682
 Uvula 622

V

Vargervik's cross-sectional study 106
 Vascular endothelial growth factor
 see VEGF
 Vascularized buccal mucosa 462
 VC 651
 VCFS (Velo-Cardio-Facial Syndrome)
 644, 647, 706, 715, 716
 Veau-Wardill-Killner procedure 640
 VEGF (vascular endothelial growth
 factor) 303
 Velar closure 632
 Velar disuse atrophy 740, 745
 Velar elevation 739, 743–746
 Velar flap 377
 Velar lengthening 640
 Velar muscular anatomy 668
 Velar section 727
 Velo-cardio-facial syndrome
 see also Vcfs 649, 682
 Velopharyngeal adequacy 607
 Velopharyngeal bulb 745, 746
 Velopharyngeal closure (s. also VPC)
 141, 573, 610, 616–618, 623–625, 627,
 684, 685, 708, 709, 722, 723
 Velopharyngeal incompetence
 see also VPC 164, 609, 707, 708
 Velopharyngeal dysfunction see VPD
 Velopharyngeal flap 739
 Velopharyngeal gap 655
 Velopharyngeal incompetence
 see also VPI 164, 175, 418, 510
 Velopharyngeal insufficiency (VPI)
 462, 515, 516, 548
 Velopharyngeal lumen 40
 Velopharyngeal management algorithm
 681
 Velopharyngeal mechanism 643
 Velopharyngeal obturation 739
 Velopharyngeal orifice area 710
 Velopharyngeal port see VP
 Velopharyngeal sphincter 685
 Velopharyngeal stimulation 739, 745,
 746
 Velopharyngeal surgery 588
 Velopharyngeal system 696
 Velopharyngeal valving 626, 628, 646,
 647, 651, 654, 669
 Velopharynx 682
 Veloplasty 698, 699
 Velum 621–624, 629, 633, 635, 643, 653,
 670, 684
 Ventral occipitotemporal cortex 711
 Ventroflexion 99, 102, 112, 129, 219, 221
 – premaxillary 112, 113
 Verlopharyngeal competence 327
 Verlopharyngeal insufficiency 64
 Vermillion border 46, 454
 Vermillion deficiency 463
 Vermillion incision 276
 Vertebral column 19
 Vertical facial growth pattern 435
 Vertical maxillary deficiency 505, 515
 Vertical maxillary excess 524
 Vertical maxillary growth 569
 Vertical prolalial tape 455
 Vicryl suture 462
 Victim of stigmatization 267, 268
 Videofluoroscopy 462, 610, 616, 652,
 653, 697
 Videonasopharyngoscopy 617, 653
 Vitronectin 302
 Vocabulary development 692, 712
 Vocal tract configuration 607
 Vocal tract resistance 664
 Voice disorder 706, 716
 Vomer 732
 Vomer flap 107, 108, 321, 327, 349, 364,
 366, 374, 415, 426
 Vomer-nasal septum complex 543
 Vomer-premaxillary suture 543
 Von Langenbeck palatal cleft
 closure 84
 Von Langenbeck palatoplasty 323
 Von Langenbeck plus vomer flap 95
 Von Langenbeck procedure 66, 68, 81,
 105, 108, 111, 118, 132, 140, 162, 179,
 194, 195, 197, 205, 305, 309, 319, 326,
 342, 351, 364, 366, 374, 375, 401, 415,
 434, 491, 698, 754
 Von Langenbeck repair 515, 516
 Von Langenbeck's technique
 see Von Langenbeck's procedure
 Von Recklinghausen's disease 5
 VP (velopharyngeal port) 608, 609,
 616, 618, 667, 672, 677
 – disorder 626
 – mislearning 626
 – movement 640
 VPC (velopharyngeal closure) 622,
 628, 633, 635, 637, 668, 670–672, 675,
 692
 – normal 643f, 647
 – postpalatoplasty 691
 VPD (velopharyngeal dysfunction)
 581, 626, 643, 667–669, 671–673,
 675–678, 681, 682, 685, 686, 705, 706,
 711, 715, 735
 – persistent 677
 – interdisciplinary team managing
 681
 – differential diagnosis 683
 – anatomic deficiency 683, 684
 – myoneural deficiency 683, 684
 – prosthetic management 684
 – surgical management 684
 – speech 705, language 705
 VPI (s. also velopharyngeal incompe-
 tency) 199, 510, 582, 608, 610, 617,
 622, 626, 635, 643, 646–651, 653–655,
 661, 664, 682, 696, 697, 706–710, 716,
 735–738, 740, 745, 746, 753
 – evaluation 650
 – indirect assessments 650
 V-Y pushback of the palate 329

V-Y pushback procedure 328
V-Y retroposition operation 320

W

Waardenburg syndrome 5
Wardill-Kilner pushback 640, 700
Wardill-Kilner V-Y pushback procedure 329
Warren/Dubois technique 619
Wassmund procedure 515
Waters' view 616, 617
Wealthy western world 237
Weight-for-length 401
WHO (World Health Organization) 770, 775

Wide alveolar cleft 547
World Health Organization see WHO
Wound contraction 301, 305
Wound healing 301
– biological mechanisms 305
– inflammatory phase 302
– mechanisms 308
– oral mucosa 301
– remodeling phase 303
– skin 301
– tissue formation phase 302

X

Xenograft 596
Xerographic study 754

Z

Zona pellucida 53, 199, 648
Zurich Approach 395, 398–400
Zurich Concept 385
Zurich treatment center 699
Zygoma-malar rotation 555